NEUROSURGERY
of Infancy And Childhood

Characteristic neurosurgical lesions found in infants and children.

A. Diastematomyelia
B. Dermoid cyst of the cauda equina
C. Subdural hematoma
D. Congenital dermal sinus expanding into intracranial dermoid cyst
E. Brain abscess associated with congenital cyanotic heart disease
F. Intracranial teratoma
G. Mid-line cerebellar astrocytoma
H. Papillary adenoma of the choroid plexus


A


B


C


D


E


F


G


H

NEUROSURGERY

of Infancy and Childhood

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RESIDENT NEUROSURGEONS

who have participated so vitally in this work during the past twenty years, and also to those whose training may in the future be facilitated by the availability of this record.

Preface

THE PURPOSE of this book is not to serve as an encyclopedic text of reference covering in detail the entire field of neurosurgery in childhood. Many of the standard disciplines of adult neurological examination and surgical therapy are equally appropriate in childhood; these will receive only cursory attention. Others differ slightly, or are entirely peculiar to the management of infants and young children. It is these differences which will be stressed.

No attempt will be made to discuss critically all of the methods that have been described to treat various neurological disorders in childhood. Neither is it our intention to imply that the methods and beliefs set forth here are the only ones acceptable or indeed necessarily always the best available. Rather, this book is a description of the diagnostic studies, principles of patient care and methods of surgical treatment which have been developed or have been found most satisfactory to date in the experience of the neurosurgical service of The Children's Medical Center, Boston.

The central nervous system in childhood, as in adults, is involved in acute and chronic trauma, purulent and aseptic inflammatory processes, toxic degeneration, and a variety of benign and malignant neoplasms. In addition, there are a wide variety of congenital malformations involving neural tissue and its coverings which are subject to surgical improvement in early life.

The importance of growth as a modifying influence on surgical physiology will be emphasized many times in the sections to follow. Because the nervous system of the child is still growing and maturing, it is possible often to carry out prophylactic surgical procedures which, though not curative of an existing lesion, may be of great significance in arresting or slowing down a progressing disturbance of function. The results of preventive surgery of this type may be unimpressive at the time of execution or in the early post-operative course, but in the long run extremely rewarding to both doctor and patient.

In the field of pediatric neurosurgery we have learned much from our colleagues in neurology, pediatrics and pediatric general surgery. It is our feeling, however, that pre-operative diagnostic studies and supportive treatment as well as post-operative care are best managed under the direction of the neurosurgeon himself. His responsibilities should not begin only at the time operation is undertaken, nor should they terminate as soon as

the surgery has been completed. Continuous care of these children under one authority is superior to division of responsibility among several.

It is hoped throughout this text not only to emphasize broad aspects of central nervous system physiology and disease which are particularly pertinent to the growing child, but also to describe various practical methods and procedures which have been found valuable to us and may perhaps prove useful to those who treat children less frequently.

* * * * *

This book is based on the experience of the neurosurgical service of The Children's Medical Center. This experience has profited from the advice of many other services and many other individuals. It does not seem appropriate or feasible here to attempt to list the large numbers of persons who have participated in this work in the past 20 years, but their interest, cooperation and tolerance are acknowledged sincerely and gratefully. Appreciation is expressed especially to the Department of Radiology under the direction of Dr. E. B. D. Neuhauser and to the Department of Pathology under the direction of Dr. Sidney Farber from whose files so much source material has been generously contributed.

In assembling this material, particular credit goes to Mrs. Nathan Laskin and Mrs. Peter Schurr for their cheerful and efficient attention to the endless details connected with preparation of the manuscript. The photographs, without which this book would be lifeless, have all been produced by the Department of Visual Education of The Children's Medical Center, under the direction of Mr. Ferdinand R. Harding. To Miss Mildred Codding and Mrs. George Homans great thanks are due for their skill and patience in creation of the original drawings. The charts are the painstaking work of Miss Edith Pierson. The photomicrographs were made for the most part by Mr. Leo Goodman of the Boston City Hospital.

Mr. Charles C Thomas and his associates have been wise and helpful counselors at every stage in the preparation and publication of this book; their cooperation, advice and insistence on high publication standards are gratefully acknowledged.

We wish to express our gratitude to those who have contributed generously to the background for this work through the Kent Research Fund.

F.D.I.
D.D.M.

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NEUROSURGERY
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PART I

CONGENITAL ANOMALIES OF THE CENTRAL NERVOUS SYSTEM AND ITS COVERINGS

Chapter I

Spina Bifida and Cranium Bifidum

Embryology

The cause of spina bifida and cranium bifidum is unknown. Whether a primary defect exists in the germ plasm which causes faulty development of the central neural axis, or whether some external agent is active after normal fertilization has occurred is of little practical significance. In the vast majority of individual patients with spina bifida or cranium bifidum, there is neither a family record of congenital abnormality nor is there a history of trauma, infection or metabolic disturbance during the first weeks of the mother's pregnancy.

The embryologic circumstances, however, which determine the type of spina bifida or cranium bifidum and the various accompanying neurological abnormalities have been studied in early human material by many investigators.[4, 21, 120, 146, 153, 185] The earliest recognizable development is differentiation of the three primary germ layers from a mass of multi-potential cells. Subsequently, at about the time of earliest segmentation, the central neural axis begins to form along the mid-dorsal aspect of the embryo. A groove appears between two longitudinally arranged mounds of cells, or neural crests. This groove deepens and the neural crests converge dorsally to form the primitive neural tube. This fusion begins at about the six-somite stage in the mid-portion of the embryo and progresses both cranially and caudally. Completion of invagination of the neural tube takes place normally in the region of the posterior neuropore at about the 30-somite stage or approximately the end of the fourth week after conception. Subsequent differentiation of the proliferating cells of this neural tube eventually results in the neuronal and supportive components of the central nervous system and perhaps part of the meninges.

During formation of the neural tube, simultaneous condensation and organization of mesodermal tissues begins to form the bone, cartilage, fat, connective tissue, blood vessels and meninges which surround and support the neural axis. Although the mechanism and extent to which ectodermal and mesodermal tissues influence one another at this stage is unkown, it is evident that the complicated differentiation of all germ layers may simultaneously go awry in single or adjacent segments. Depending upon which

primitive tissues are involved and to what degree, a vast variety of clinical malformations becomes possible, from the simplest type of asymptomatic occult laminar defect in the lumbar spine to complete spinal or cranial rachischisis.

Although the mesodermal defect which results in spina bifida and cranium bifidum is undoubtedly initiated in the first few weeks of embryonic life, the clinical form of the lesion is influenced by further changes in the relationship of the central neural axis to surrounding structures as growth proceeds. During the second month of gestation, the most caudal vertebral segments fuse and the corresponding neural elements become vestigial. From the third month on, the vertebral column together with its associated mesodermal structures, fascia, fat, cartilage, and meninges, grows at a more rapid rate than the spinal cord itself. Because the position of the spinal cord is fixed cranially due to its attachment to the brain stem within the cranial vault, the greatest change in its relation to corresponding vertebral segments occurs caudally. As long as this differential rate of growth continues, the spinal cord appears to migrate cranially and the more rapidly growing vertebrae necessarily shift caudally. Whereas the spinal cord and vertebral column are of equal length at the third month of embryonic life, by the time of birth the conus medullaris is found opposite the third or fourth lumbar vertebra and by the time growth has ceased, it lies opposite the first lumbar vertebra or slightly cranial to this point. The clinical significance of this differential growth rate will be elaborated in subsequent sections of this chapter.

The following discussion is based directly on the study and treatment of over 1,400 patients with spina bifida by the neurosurgical service of The Children's Medical Center. The incidence of the various types of spina bifida and cranium bifidum in this series is shown in Table I.

TABLE I

INCIDENCE OF TYPES OF SPINA BIFIDA AND CRANIUM BIFIDUM

Type of Lesion	*No. of Cases*
Spina bifida occulta (hospitalized cases only)	96
Meningocele	396
Myelomeningocele	754
Encephalocele	187
Total	1,433

SPINA BIFIDA OCCULTA

In ordinary usage, the term spina bifida occulta includes all lesions in which fusion defects of the vertebral column are present without protrusion of intraspinal contents to the surface. This is perhaps the commonest of all

congenital "anomalies." Probably about 25 per cent of all children will show some minor defect of a vertebral spine or lamina on roentgen examination. Most of these occur in the lumbo-sacral area and persist into adult life in normal individuals who show no evidence of either neurological or musculo-skeletal deficiency. By far the majority of occult spinal disorders, therefore, are of no clinical significance. There is, however, a comparatively small group in whom the central neural axis is involved as well as the bone and in whom surgical therapy in early life may be beneficial.[102]

Since the primitive mesoderm which provides the ground substance for the bones of the spinal axis also gives origin to fat, fibrous tissue, corium and blood vessels, it is not surprising to find a variety of abnormalities of these structures occurring in association with bony defects. As already pointed out, growth and development of the nervous system continue after birth and there is a constantly changing anatomical relationship between the spinal cord and surrounding structures as growth proceeds. It is important, therefore, to detect that group of patients with occult spinal defects who show evidence of neurological disorders which may be subject to relief or arrest of progression by surgical treatment. Since the differential rate of growth of the spinal cord and spinal column becomes gradually less as childhood progresses, it is desirable to recognize these patients as early in life as possible.

Symptoms and Signs

Aside from those patients in whom the diagnosis of spina bifida occulta is made incidentally on x-ray films of the spine made for some other purpose, these lesions are usually detected clinically for one of two reasons. Either a cutaneous or subcutaneous abnormality points to an underlying spinal defect, or else investigation of a neurological disorder, particularly of the lower extremities, bladder or bowel, leads to a diagnosis on subsequent x-ray examination.

Cutaneous Defects: Four different types of cutaneous or subcutaneous abnormalities are commonly seen associated with occult spinal disorders. These may be of sufficient size or disfigurement to constitute the patient's primary complaint or they may be picked up in the course of routine physical examination. They may occur singly or in various combinations.

(1) Abnormal Hair: An unusual growth of hair may be present at birth; this is usually maximal in the mid-line, tapering off laterally. Such hair may be coarse and several inches in length, even in early infancy, or it may be silky down limited to a discrete area (Figure 1).

(2) Angiomas: Cutaneous "port wine" stains of irregular size and distribution are present in a large proportion of children in the suboccipital

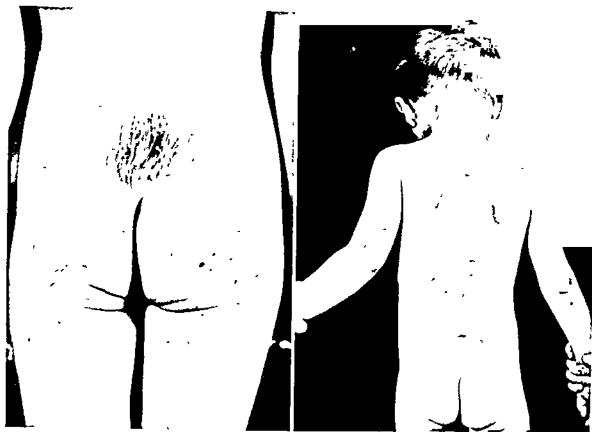


Figure 1 (*Left*). Five and four-twelfths year old girl with spina bifida occulta showing abnormal tuft of hair in the lumbo-sacral region. (Reprinted through the courtesy of The Massachusetts Medical Society from *New England J. Med.*, 228.745-750, 1943.)

Figure 2 (*Right*). Angiomatous cutaneous malformation in the lumbar region associated with spina bifida occulta and an intraspinal dermoid cyst at the upper lumbar level in a 2 8/12 year old boy.

area; in this location they are of no clinical significance. However, when these superficial angiomas are present in the mid-line along the spine, and particularly in the lumbo-sacral area, they are frequently associated with an underlying spina bifida (Figures 2 and 5).

(3) Lipomas: Subcutaneous, non-tender, poorly circumscribed, soft, partly movable masses of fat are very commonly associated with occult spinal defects. These occur in the mid-line, or just off the mid-line extending toward it, and may vary in size from poorly visualized but palpable nodules a centimeter or two in diameter up to huge bustle-like protrusions 10 or 12 cms. across. The overlying skin may be smooth and normal in appearance or marked by dimpling, abnormal hair or abnormal vascularity. These lipomatous masses consist of lobules of a firm gritty type of fat bound together with numerous fibrous septa and usually adherent to fascial or dural planes. They may be located at any level from the subcutaneous tissues down to the cord itself, as will be emphasized later (Figures 3 and 4).

(4) Dimples: Depressions in the skin in the mid-line or just off the mid-

line with fixation of the epithelium to underlying layers should also alert the observer to an underlying *spina bifida*. *Such dimples may be minute* and discovered only on careful examination, particularly of the intergluteal cleft, or they may present as open sinus tracts with inflammatory reaction and scarring of the surrounding skin (Figures 3 and 5). These cutaneous dimples may simply mark the level of an underlying occult spina bifida or they may mark the outlet of a continuous fibrous or fistulous tract extending directly into the spinal canal as will be discussed in the chapter on Congenital Dermal Sinus Tracts (p. 69).



Figure 3 (*Left*) Dimple to the left of the mid-line and subcutaneous fat pad about 4 cms in diameter in a 2 8/12 year old child with spina bifida occulta and intraspinal lipomeningocele.

Figure 4 (*Right*). Three year old girl with spina bifida occulta and large extraspinal mass consisting of fat and anomalous bone.

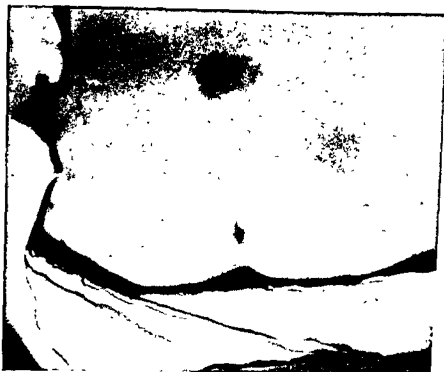




Figure 5 (*Upper*). Lumbo-sacral region of infant showing area of telangiectasis and mid-line dimple which led through area of spina bifida to intraspinal dermoid cyst. (Reprinted through the courtesy of The Yorke Publishing Company Inc. from *Am. J. Surg.*, 75 231-236, 1948.[7])

Figure 6 (*Lower*). (*See legend on facing page.*)



Figure 7. Intravenous urogram of patient with spina bifida occulta with large lumbo-sacral intraspinal lipomeningocele. Note unilateral hydronephrosis and marked atonicity of the bladder. Patient was incontinent of urine.

Neurological Disorders: From a surgical point of view, the patients who are discovered to have spina bifida occulta in the course of investigation of a neurological deficit are of particular importance. This group of patients can be divided usefully into two sub-groups; those who complain of musculo-skeletal or sensory disorders of the lower extremities and those whose complaints relate to control of the bladder and rectal sphincters.

(1) *Musculo-skeletal Disorders:* These patients are often discovered at about the beginning of the second year of life as they are, or should be,

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Figure 6 (*Lower*). Marked deformity of the left foot associated with spina bifida occulta of the lumbar region with intra- and extraspinal lipoma. (Reprinted through the courtesy of The Massachusetts Medical Society from *New England J. Med*, 228:745-750, 1943.)

making their first attempts to walk. Muscle weakness, deformities of the feet and hips and gait disturbances are first noted at this time by the family doctor or pediatrician. Roentgen examination, often after orthopedic consultation, discloses the existence of spina bifida occulta. Valgus, varus and cavus deformities of the feet, atrophy of the lower leg musculature, limp, deformities of the toes, real or apparent shortening of one leg, pelvic tilt and scoliosis have all been noted as the initial complaint (Figures 6 and 8). Defects of the vertebral bodies and pedicles are often seen with spina bifida, and the presence of associated hemivertebrae or fusion of two or more vertebrae may produce a variety of structural deformities. Numerous abnormalities of posture and gait are noted, depending upon which muscle groups show weakness. Every child, therefore, who shows a congenital type of scoliosis, dislocation of the hips, pelvic asymmetry, positional deformity of the feet, absent deep tendon reflexes or evidence of sensory impairment in the lower extremities should have careful roentgenograms made of the spine.

(2) Sphincter Disturbance: These abnormalities in minor degree are often overlooked in early infancy and discovered only at the time the child should ordinarily be trained. Especially significant are: (1) regression of bladder and bowel control or training habits once learned; (2) actual demonstration of urinary dribbling in response to abdominal pressure; and (3) palpable relaxation of the anal sphincter. *Spina bifida is often*



Figure 8. Two and three-twelfths year old girl with spina bifida occulta accompanied by urinary incontinence and the visible deformities of the lower extremities. Myelogram and operation revealed a lipoma of the cauda equina.

disclosed on x-ray films made during intravenous urography in the study of poor urinary sphincter control (Figures 7 and 8). Certainly when sphincter disturbance persists in the absence of local pathology, the significance of an occult defect in the lumbo-sacral spine should be carefully evaluated.

X-ray Examination

Because of the thinness of the laminae and absence of ossification in early infancy it is often difficult to detect minor degrees of spina bifida on plain x-ray films, particularly in the cervical and lumbo-sacral areas where the canal is normally widened. Otherwise, good anterior-posterior films of the spine are usually satisfactory to detect the extent of any fusion



Figure 9. Three years and three months old girl with spina
anomalies of the
that patient has

defect, abnormal widening of the spinal canal, anomalies of the vertebral bodies such as single or double hemivertebrae or fused vertebrae, or the presence of bony spicules or other anomalous bony masses in the vertebral defect (Figures 9 and 10).

Opaque myelography is an invaluable adjunct in evaluation of occult spinal disorders. With pantopaque this is a safe and easily accomplished diagnostic procedure at any age (Figure 11). In uncooperative infants and younger children general anesthesia is often necessary, but ordinarily mild sedation and local procaine are sufficient. Under fluoroscopy the width

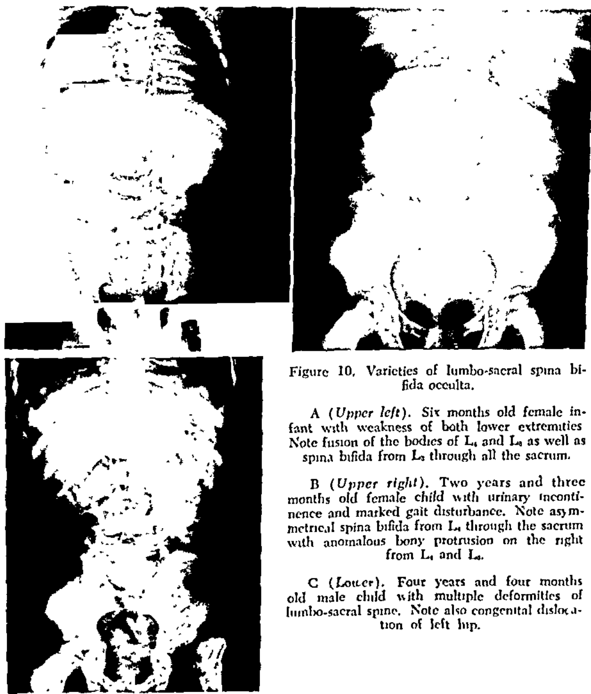


Figure 10. Varieties of lumbo-sacral spina bifida occulta.

A (*Upper left*). Six months old female infant with weakness of both lower extremities Note fusion of the bodies of L_4 and L_5 as well as spina bifida from L_5 through all the sacrum.

B (*Upper right*). Two years and three months old female child with urinary incontinence and marked gait disturbance. Note asymmetrical spina bifida from L_4 through the sacrum with anomalous bony protrusion on the right from L_4 and L_5.

C (*Lower*). Four years and four months old male child with multiple deformities of lumbo-sacral spine. Note also congenital dislocation of left hip.

of the dural canal at any level is actually visualized and obstruction to or deformities of the flow of the opaque medium in the subarachnoid space become apparent immediately. Filling defects caused by intraspinal lipomatous masses, fibrous cords, bony spicules, dermoid cysts, intraspinal meningoceles or other lesions are readily detected and localized. Spot films made at the time record these abnormalities (Figures 12 and 13).

Indications for Surgery

Surgical exploration in spina bifida occulta should be performed for specific indications. If severe neurological deficit is present at birth there is probably little to be gained by surgical treatment. However, if evidence of neurological abnormality is first noted at some time after birth, then it may be assumed reasonably that continued growth may increase the severity of the defect. Likewise, if under observation musculoskeletal abnormality or sphincter disturbance progresses, an attempt should be made to halt this process.

A natural tendency to delay radical exploration can frequently be resolved by myelography. If pantopaque myelography demonstrates a filling defect in the region of spina bifida in a patient who has clinical evidence of impaired neurological function, then surgical exploration is warranted. This should be regarded more often as prophylactic than curative surgery. The primary purpose of operation is to restore conditions structurally to as nearly normal as possible in an attempt to prevent an increase in the neurological deficit with further growth.



Figure 11. Two normal pantopaque myelograms in infants under six months of age.

The attitude toward surgical therapy should be one of early exploration for the indications as outlined, but conservative treatment once surgery has been decided upon. There is certainly little to gain, and there may be much harm done by injudicious manipulation of the spinal cord and cauda equina.



Figure 12 (*Left*). Typical myelogram often seen in patients with occult spina bifida showing marked widening of the canal, but in this case no evidence of an intraspinal meningocele or mass and therefore a poor candidate for improvement following surgical exploration.

Figure 13 (*Right*). Pantopaque myelogram showing opaque medium in the lumen of a lipomeningocele in patient with spina bifida occulta and progressive gait disturbance.

In so far as mass lesions can be removed safely, fibrous bands divided, bony spicules excised, and adhesions of neural structures or of meninges to the surrounding soft tissues separated, surgery may be of considerable benefit. *Attempts to dissect free or to transplant the actual position of the cord or the roots of the cauda equina are almost certain to result in an increased neurological deficit.*

Operation

Exploration is carried out as any formal laminectomy in childhood, with the patient in the prone position under general inhalation anesthesia. It is important to remember that anomalies may be found as soon as the midline skin incision is made. Since the meninges may be defective as well as the bone and subcutaneous structures, nerve tissue may be encountered at any stage of the exploration. It is usually wise, therefore, to expose a normal bony segment cranial or caudal to the region of the spina bifida. Laminectomy is then commenced where the structures are in normal relationship and carried toward the abnormal area.

Operative findings vary considerably, and no two cases are apt to be

exactly similar. The margins of defective laminae are identified and removed as necessary to facilitate intraspinal exploration. Cartilaginous nubbins are often found in the region of the bony gap and should ordinarily be removed. As already mentioned, poorly circumscribed masses of anomalous fatty tissue may be found in any layer from the subcutaneous tissue to the intrathecal space. This fat may be continuous from extraspinal to intraspinal compartments. It is particularly important not to confuse nerve roots with the fibrous septa so commonly found traversing this anomalous lipomatous tissue. Electrical stimulation is often helpful to this end as the excision of intraspinal fatty tissue proceeds. It is usually unwise to attempt complete excision of lipomatous masses which extend intradurally because of their extremely close adherence to the spinal cord itself or to the roots of the cauda equina. However, the entire neural axis and its covering meninges should be freed within the bony canal from adherence to extraspinal tissues. If the filum terminale is thickened and under tension it should be divided.

When a fibrous tract or stalk extends from the skin directly into the dura, the latter should be widely exposed and opened to rule out the presence of an intradural dermoid cyst, lipoma or adherent fibrous septum through the cord. Such fibrous tracts should always be excised. The management of bony spicules dividing the spinal contents into two compartments will be discussed in the section on Diastematomyelia (p. 57).

Occasionally an intraspinal meningocele is present in occult spina bifida (Figure 14). A definite sac may extend up or down inside the bony canal or, more commonly, mushroom out in the soft tissues after protruding through the bone defect by a narrow stalk. In this situation, the meningocele sac should first be mobilized carefully, then opened and its contents inspected. If there are no neural elements the neck of the sac is amputated at the dural level. If neural elements are attached to the walls of the meningocele, they are freed up if possible or if not, that portion of the meningocele sac is left behind as described in the section on meningocele repair (p. 35).

Following exploration in spina bifida occulta it is usually wise to leave as wide a decompression as possible. Therefore, no attempt is made to repair the bony defect, or to swing fascial flaps across the defect as in meningocele repair. The wound is closed in layers in the usual fashion although frequently the paraspinal muscles are so far separated and defective that they cannot be approximated very satisfactorily.

Material

In our clinic 96 patients with spina bifida occulta have been admitted to the hospital for observation and treatment. Thirty-nine of these presented

because of a cutaneous abnormality, 45 because of a neurological deficit in the lower extremities, and in 19 the initial complaint was related to control of the rectal or vesical sphincters (Table II).

Table II

SPINA BIFIDA OCCULTA
CHIEF COMPLAINT OF HOSPITALIZED CASES

	No. of Cases
Cutaneous Abnormality	39
Lower Extremity Deficit	45
Sphincter Disturbance	19

The distribution of the level of spina bifida occulta in these cases is shown in Table III.

Table III

SPINA BIFIDA OCCULTA
CASES HOSPITALIZED FOR STUDY

Level	*No. of Cases*
Cervical	4
Thoracic	7
Thoraco-lumbar	8
Lumbar	22
Lumbo-sacral	34
Sacral	21
Total	96

In this group of patients, 13 have had myelography performed. In six of these a demonstrable deformity was present and in seven myelography was negative. Forty-four patients had surgical exploration of the region of the spina bifida. The findings at operation are shown in Table IV.*

It is extremely difficult to evaluate the efficacy of surgical treatment in any single patient because this is primarily prophylactic rather than cura-

Table IV

SPINA BIFIDA OCCULTA
OPERATIVE FINDINGS

Total Patients Operated on		44
Findings at Operation		
Lipoma	23	
Stalk	15	
Intraspinal Meningocele	3	
Dermoid Cyst	1	
No Intraspinal Abnormalities Found	8	

* *Diastematomyelia and congenital dermal sinus tracts are not included in this group.* *See* p. 57 and p. 69

tive surgery. It is impossible to state accurately how far a neurological deficit might have progressed had not an attempt been made to delay or arrest it. There has been no operative mortality in this series. With careful surgery there should be little risk of increasing neurological defects, except perhaps temporarily. There have been sufficient follow-up observations to conclude that in those patients in whom some actual structural abnormality was found, there has been improvement or arrest of progressive neurological involvement in a high percentage of instances.

SPINAL MENINGOCELE AND MYELOMENINGOCELE

From a neurosurgical point of view, the most important aspect of spina bifida is the treatment of patients with various types of meningocele and myelomeningocele. These are discussed together because there is often no reliable clinical differentiation between them and because they present related surgical problems.

In general, if there is a visible sac along the spinal axis which is covered with epithelium or with a membrane in which no nerve tissue can be

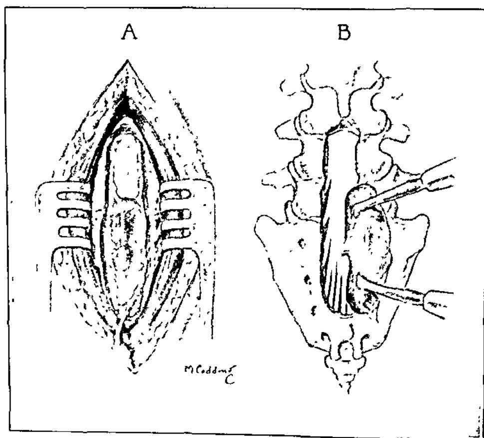


Figure 14 These drawings show an intraspinal meningocele connected through the dura by a small stalk in a patient with spina bifida occulta. (Reprinted through the courtesy of The Massachusetts Medical Society from *New England J. Med*, 228:745-750, 1943.)

seen directly or by transillumination, and if on neurological examination there is no evidence of sensory, motor or reflex abnormality or of sphincter disturbance, the lesion is termed a meningocele. If nerve tissue can be seen within or adherent to the sac, or there is a clinically demonstrable neurological deficit caudal to the level of the lesion, it is termed a myelomeningocele. It is obvious that in new-born infants, finer abnormalities of motor, sensory, reflex and sphincter function may be difficult or impos-

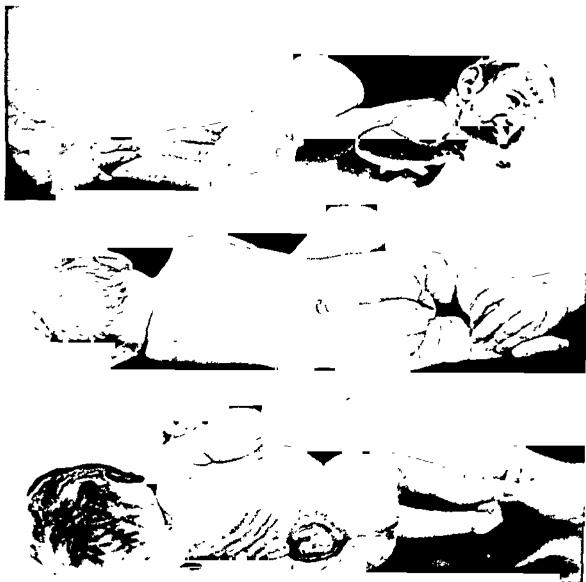


Figure 15 (*Top*). Huge neuro-enteric defect associated with lumbo-sacral spina bifida. Part of this mass represented a meningocele sac containing clear fluid and the rest a teratoid tumor.

Figure 16 (*Middle*). Six months old infant with flat membranous meningocele containing no neural tissue. Narrow meningeal stalk with patent lumen extended to the surface.

Figure 17 (*Bottom*). New-born infant with lumbo-sacral spinal rachischisis showing large area of exposed neural tissue. Complete paralysis of lower extremities and sphincters. No treatment indicated.

sible to assess. It is also obvious that in many lesions it is impossible to be certain by direct inspection of the surface and by transillumination whether or not neural elements may be present. Frequently, therefore, the designation may be quite arbitrary until operative or pathological data are available. As will be noted from Table I, the incidence of myelomeningocele is about twice that of meningocele in this clinic.

The diagnosis of meningocele is ordinarily self-evident at the time of birth. Occasionally, there may be difficulty in differentiating a caudally situated lesion from a sacro-coccygeal teratoma. This can usually be resolved by plain roentgenograms of the spine or by appropriate lateral roentgenograms after injection of a small amount of air into the spinal subarachnoid space. The two lesions may be seen in combination (Figure 15).

From an anatomical point of view, there is an infinite variety of lesions, varying from the simplest flat, partially epithelialized membrane overlying a bony defect (Figure 16) to complete spinal rachischisis in which most of the spinal cord may present directly on the surface (Figure 17). If a meningocele sac protrudes beyond the skin surface, as is the rule, a pedunculated lesion with a broad sac on a narrow stalk-like base may result (Figure 18), or more commonly, a so-called sessile type of lesion occurs, in which the sac is widest at the level of the skin surface (Figure 19). The covering of a meningocele sac may be entirely membranous, transparent, thin, and prone to easy ulceration and rupture from external friction; it may be translucent, thickened, irregularly scarred, and partially covered by vascular malformation (Figure 20); it may be partially or totally covered with epithelium or with full-thickness skin (Figure 31). Associated with many meningocele and myelomeningocele sacs, as with spina bifida occulta, there is often an overgrowth of fatty tissue. This is a peculiar gritty type of fat, crossed by many fibrous tissue strands usually closely adherent to the meningocele sac (Figures 21 and 22).



Figure 18. Pedunculated skin-covered upper dorsal meningocele in new-born infant with no neurological abnormalities demonstrable.

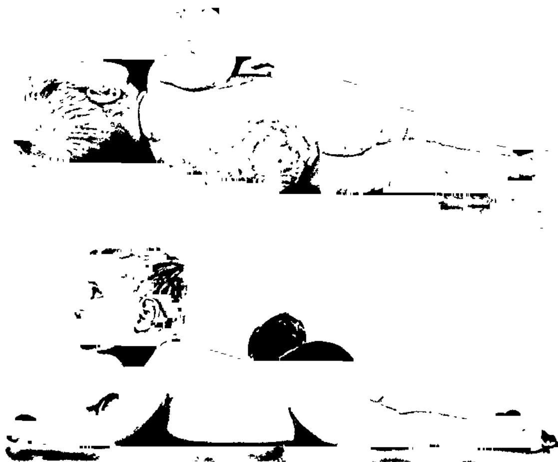


Figure 19. Huge, sessile type of lumbo-sacral myelomeningocele, partially covered with skin. Patient has paraplegia but no evidence of hydrocephalus.

These lesions may occur at any point along the spinal axis but are seen with greatest frequency in the lumbo-sacral area. The next most common area is the cervical spine, and a much smaller percentage are scattered along the low cervical, thoracic and upper lumbar regions. Anterior and lateral meningoceles are rare lesions which should be kept in mind in the differential diagnosis of atypical masses in the thorax, retroperitoneal tissues and pelvis (Figures 23, 24 and 25). The importance of correct diagnosis, usually by myelography, previous to operative treatment in these instances is obvious. The anatomical distribution of cases in our series in seen in Table. V.

Physical examination of the patient with spinal meningocele should include: (1) careful inspection of the local lesion; (2) neurological examination of the extremities and sphincters; (3) evaluation of the presence of hydrocephalus; and (4) check for other congenital anomalies.

(1) Local Lesion: Attention is directed to the size of the lesion particularly at its junction with the skin surface and to the character of the surrounding skin. Measurements are made and recorded. Careful inspection of the surface for neural elements and for areas of ulceration and leak-

age is made. The lesion should then be transilluminated with a strong light in a darkened room to disclose the possible presence of nerve tissue within the sac. Careful palpation will usually reveal the extent of spina bifida with reasonable accuracy.

TABLE V

MENINGOCELE AND MYELOMENINGOCELE
SITE OF LESION

Level		*No. of Cases*
Cervical		47
Thoracic		82
Thoraco-lumbar		133
Lumbar		457
Lumbo-sacral		328
Sacral		97
Anterior		4
Thoracic	2	
Pelvic	2	
Undesignated		9
Total		1,157

(2) *Neurological Examination:* In the new-born period and in early infancy the limitations of neurological examination are many. However, gross neurological deficit in the lower extremities is readily apparent. Paralysis in lumbo-sacral lesions is ordinarily of the flaccid variety and in severe degrees of myelomeningocele is apt to be complete from birth. Associated with complete loss of voluntary motion there is total absence of sensation and total areflexia. The feet and legs under such circumstances may or may not be deformed at birth. In cervical and thoracic lesions with partial involvement of the spinal cord, spasticity of the lower extremities with hyperactive deep tendon reflexes, ankle clonus, and extensor plantar responses may be seen.

All varieties of muscular imbalance in the lower extremities may occur depending upon which elements of the cauda equina are involved. As a result flexion contracture or dislocation of the hips and equino-varus and valgus deformities of the feet are seen frequently, as well as less striking abnormalities of the toes and forefoot (Figure 26).

Relaxation of the rectal sphincter may be apparent even in the new-born (Figure 26). Eversion and fissuring of the anal skin is seen and there is no resistance to introduction of the examining finger. The anal skin reflex is absent. Less severe degrees of rectal incompetence are extremely difficult to assess in early life.

Constant dribbling of urine accentuated by any suprapubic pressure or handling of the patient implies loss of innervation to the vesical sphincters.

This lack of urinary control is usually indicated by early maceration of the perineal skin. Partial degrees of both vesical and rectal sphincter disturbance may improve remarkably with training as a child becomes able and anxious to learn.

(3) Hydrocephalus: The standard occipito-bregmatic circumference should be measured with a steel tape and compared with the size of the chest and with standard charts (Figures 27 and 28). This measurement should be made at regular intervals on every meningocele patient being followed, since the rate of growth is more important than any one measure-



Figure 20 (*Upper*). Lumbar meningocele with retracted, thickened, partially epithelialized surface. Negative neurological examination. Favorable for early resection.

Figure 21 (*Lower*) Soft lipomeningocele completely covered with full thickness skin. Partial paraplegia and weakness of anal sphincter

ment. The ordinary signs of hydrocephalus in infancy are all sought for; dilatation of the scalp veins, shinyness of the scalp, tension of the fontanelle, separation of the cranial sutures, hollowness of the cranial percussion note, downward displacement of the eyes. The presence of hydrocephalus is the single, most discouraging factor in the management of patients with spina bifida. It is most common, of course, in patients with myelomeningocele and is the result of the Arnold-Chiari malformation, as will be pointed out subsequently (p. 42).

(4) Congenital Anomalies: The incidence of congenital disorders elsewhere in the body is higher in these patients than among the general population. The examiner should therefore be alert to the possibility of congenital heart disease, intestinal malformation, cleft palate, craniosynostosis, umbilical and inguinal hernia, congenital dislocation of the hips, clubfoot, skeletal deformities such as the Klippel-Feil syndrome and Sprengel's deformity, pilonidal sinus, and genito-urinary anomalies, all of which have been seen in this clinic.

Indications for Operation

Perhaps the most controversial aspect of the management of meningoceles and myelomeningoceles is determination of the indications for sur-



Figure 22A (*Upper*). Large lipomeningocele completely covered with skin in partially paraplegic infant.

B (*Lower*). No evidence of hydrocephalus at any time before or after excision of lesion.

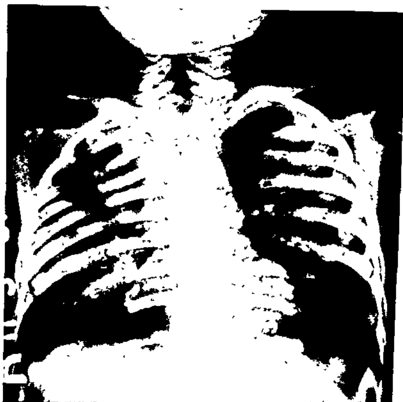


Figure 23. Antero-posterior roentgenogram of the spine of a 4 4/12 year old boy who entered the hospital complaining of wheezing and chronic bronchitis. Note spina bifida of the low cervical and upper thoracic spine with fusion of the bodies between T_2 and T_6. There is a large mid-line defect through the bodies anteriorly and a soft tissue mass extending into the right upper chest. This proved to be a neuro-enteric cyst containing intestinal epithelium connected by a narrow stalk about 5 mms. in diameter passing through the bony defect and attached to the ventral aspect of the spinal cord in the mid-line. Treated by division of the stalk and closure of the dural defect at laminectomy and removal of the cyst subsequently at thoractomy.

gical repair and the optimum time this should be performed. There are few clear-cut rules which can be enunciated; each patient must be evaluated individually. The various factors which enter into the decision include: the age and general condition of the patient, the size of the sac and particularly the width of its base, the condition of the skin surrounding the lesion, the presence or imminence of ulceration or frank leakage from the sac, the presence of arrested or progressive hydrocephalus, the neurological status of the lower extremities and sphincters, the co-existence of other anomalies and even the domestic and economic problems of the family.

In general, every mass lesion should be removed as soon as it is technically feasible and there is reasonable certainty that a useful citizen capable of self-sustaining development can be salvaged. There is seldom necessity



Figure 24 Barium enema showing displacement of large bowel by *anterior meningocele in the pelvis.* (Reprinted through the courtesy of The Massachusetts Medical Society from *New England J Med.,* 228 631-641, 1943)

for operation in the post-natal period. However, it is frequently desirable when the covering membrane is thin, ulcerated or ruptured but uninfected, the lesion is small enough to make technical repair feasible, the head is normal in size, and there is no apparent neurological deficit, to carry out repair in the first few days of life. This is particularly true in cervical and thoracic lesions. Surgery in this age period is well tolerated under local anesthesia.

It is usually preferable to delay operation for several reasons: (1) the development of some regularity of bowel movements makes wound contamination less likely; (2) the skin surrounding the meningocele base becomes toughened; (3) an estimate of the rate of head growth can be made as well as (4) a more reliable appraisal of the neurological status of the lower extremities.

In this clinic it is felt that surgery is contra-indicated in early life, even if spinal fluid leak exists, in the presence of total paralysis of both lower extremities and complete loss of rectal and vesical sphincter control (Figures 17 and 26). This is true whether or not hydrocephalus is present at the time of examination. We have not felt it justifiable or desirable to put such infants through surgical procedures, and to put parents through such added expense and heartaches where it has been clear that the patient was hopelessly and permanently crippled and that the chances of surviving to adult life in a useful fashion without encountering disastrous infection or gross



Figure 25. Antero-posterior roentgenogram of the lumbo-sacral spine showing smooth circular defect through the bony sacrum. Patient had a grapefruit-sized anterior meningocele in the pelvis which had caused chronic ureteral and large bowel obstruction. Treatment by removal of the meningocele at pelvic laparotomy and closure of the dural defect with reinforcement of the bony defect by tantalum gauze at subsequent laminectomy.

mental retardation from progressive hydrocephalus were so limited. It is obvious to state here, but by no means obvious to many parents, that elements of the central nervous system which have never developed or are once destroyed never replace themselves, and the removal of a myelomeningocele sac does not bring about recovery of paralyzed limbs or sphincters.

If, on the other hand, patients with paraplegia survive on conservative therapy, do not develop infection or progressive hydrocephalus, and appear to be developing mentally so that it is obvious they are going to be teachable, then excision of the myelomeningocele as a cosmetic procedure and as a means of facilitating nursing care and paraplegic ambulation should be carried out. This type of surgery is usually performed in children anywhere from 18 months on up to 10 or 12 years of age.

Early surgery is indicated in the presence of partial paralysis in the lower extremities and partial disturbance of the sphincters in the absence of hydrocephalus if it is apparent that enough function may be present to permit the child to be trainable and ambulatory. Patients who are denied surgery because of early hydrocephalus or because of incomplete neurological loss should be followed carefully because occasionally a situation which looks almost hopeless in early life will progress to a point where operation becomes both feasible and advisable. Removal of the myelomeningocele is contra-indicated in the presence of rapidly progressive hydrocephalus, but may become justified if the hydrocephalus arrests spon-



Figure 26. New-born infant with large sessile, lumbo-sacral myelomeningocele. Note the fetal position of the deformed lower extremities and the rectal prolapse. No surgical treatment indicated. (Reprinted through the courtesy of The Yorke Publishing Company, Inc. from *Am. J. Surg.*, 75:231-256, 1948.[7])

taneously or can be arrested by suboccipital craniectomy or other measures, as discussed in the section on the Arnold-Chiari malformation (p. 42).

Even with modern antibiotic protection, it is unwise to operate in the presence of extensive superficial ulceration and infection of the meningocele or of dermatitis of the surrounding skin. The organisms involved in infected meningoceles are usually *b. coli* and *staph. aureus.*

Pedunculated meningoceles can usually be removed at any age as far as technical problems are concerned. However, it is unwise to attempt excision of a sessile lesion too early if further delay, permitting epithelializa-

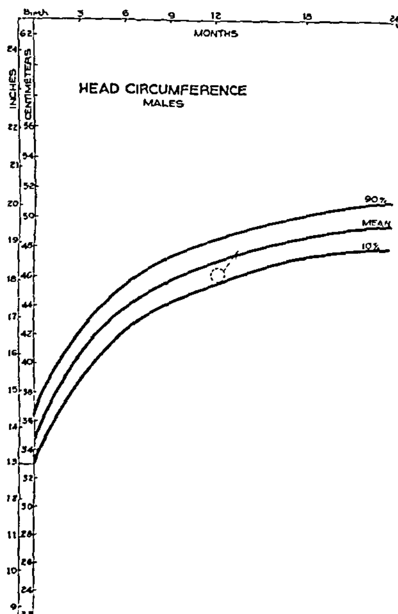


Figure 27. Chart indicating normal range of increase in head circumference in male infants during first two years of life (Reprinted through the courtesy of *The C. V. Mosby Company* from *J. Pediat.*, 33:167-171, 1948.)

tion of the sac around the base and relative decrease in the size of the lesion compared to the size of the child, will facilitate later removal without undue tension of the repair. A delay of many months is often indicated to increase the possibilities of an optimum technical result. Such delay, of course, implies that the covering of the meningocele is sufficiently tough to make conservative home management safe. In the presence of a very thin membrane one must accept the added risk and technical difficulties of early repair in order to prevent leakage of spinal fluid and meningeal infection. Such "early" repair is usually performed somewhere from the third

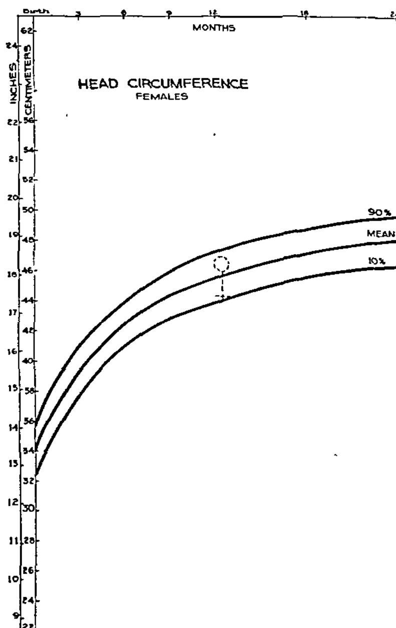


Figure 28. Chart indicating normal range of increase in head circumference in female infants during the first two years of life. (Reprinted through the courtesy of The C. V. Mosby Company from *J. Pediat.*, 33:161-171, 1948.[10])

to eighth week of life. In the event of meningitis, surgery should be deferred until clinical, bacteriological and cytological evidence of infection has disappeared. In general, however, if operation is delayed until the infant is between three and 12 months of age, time will be sufficient for development and recognition of neurological disabilities, for detection of hydrocephalus, for establishment of some regularity of bowel and bladder habits, for local growth of skin to facilitate more adequate surgical repair, and for the child to develop into a better operative risk.

Home Treatment

If operative repair of a spinal meningocele or myelomeningocele is delayed because the size of the sac obviates satisfactory closure of the skin or because it is desirable to evaluate more carefully the extent of neurological deficit in the lower extremities or the rate of head growth, parents can be taught to take care of the infant at home with little difficulty. It is neither necessary nor desirable for such infants to remain hospitalized until surgery can be performed. Similar instructions are given to parents who elect to administer terminal care at home and to patients in whom surgical repair is contra-indicated because of extensive or rapidly progressive hydrocephalus.

The home treatment of these infants is directed toward: (1) dressing the lesion itself; (2) care of the surrounding skin; (3) physiotherapy to the lower extremities; and (4) observation for evidence of hydrocephalus.

(1) *Meningocele Dressings:* It is well to show the mother or nurse how to dress a meningocele before the child leaves the hospital and then to have her do the dressing herself under observation (Figure 29). It is also our practice to give the parents a mimeographed sheet of simple instructions to which they may refer at home as necessary. These instructions together with reassurance that when the lesion is properly dressed there is little or no danger of its rupturing when the baby is handled or moves about and reassurance that the lesion is not painful when adequately protected are invariably greatly appreciated by anxious parents. The instructions are as follows:

Equipment

1. Two small basins (cereal bowls) to hold boric acid solution and alcohol
2. Sterile gauze squares, three by three inches (Drug Store).
3. Cilkloid (perforated plastic film) sections, cut one-half inch larger than meningocele (Medical Supply House)
4. Boric acid solution. (One teaspoon boric acid powder to one pint boiled water.)
5. Rubbing alcohol (Drug Store).

6. Doughnut-shaped ring of cotton or foamrubber wrapped firmly with two-inch gauze bandage to fit and protect meningocele. (Unsterile rolls of Grade B cotton can be purchased at any Drug Store.)

Procedure

1. Wash meningocele and surrounding area with boric acid solution. Be careful not to rub surface roughly.



Figure 29. Method of dressing meningocele during home treatment.

A Necessary materials: two clean basins, one with alcohol and the other with boric acid solution, sterile gauze squares, absorbent cotton, *three-inch* gauze bandages, flannel binder and safety pins.

B Absorbent cotton is made into a "doughnut" and wrapped with gauze bandage. "Doughnut" should be sufficiently large so that meningocele is *not touched* on any aspect.

C. Plastic film is placed over the meningocele.

D After covering the plastic film with sterile gauze squares, "doughnut" is put into position

E. Flannel binder holds the "doughnut" in place and baby can be handled or turned spontaneously without danger.

2. Take square of cilkloid out of alcohol, rinse it in boric acid solution and cover the meningocele.
3. Put dry sterile gauze squares over the cilkloid.
4. Place cotton doughnut around meningocele.
5. Hold doughnut in place with binder or piece of flannel and fasten firmly with safety pins.

Things to Remember

1. Procedure is clean but not sterile.
2. Wash your own hands for two minutes in soap and water before starting the dressing.
3. Boil bowls for 10 minutes before using.
4. Soak cilkloid square beneath surface of alcohol for one hour before using. Be sure to rinse it in boric acid solution before putting it on the meningocele.
5. Cilkloid need be changed only about once a week unless it becomes soiled. The sterile gauze should be changed one or more times daily.
6. If there is leakage of clear fluid from the meningocele or local evidence of infection, call your physician.

(2) *Skin Care:* Because of incompetence of the rectal and vesical sphincters in many patients with lumbo-sacral myelomeningoceles, care of the skin of the perineum, buttocks, and inner aspects of the thighs often becomes a major problem. Diapers should be changed more frequently than normal and the skin washed and dried carefully several times a day. Areas of abrasion or frank ulceration should be well covered with a bland ointment at all times. Frequent exposure to sunlight is desirable and parents are urged to leave youngsters without diapers pinned on to them as much as possible.

(3) *Physiotherapy:* If there is any impairment of function of the lower extremities, parents are instructed to carry out daily massage and passive activity at home. All joints should be carried through a full range of motion several times daily. During play periods or after feedings this can readily be done by the mother with occasional check-ups and instruction by a trained physiotherapist. When surgical repair of spina bifida is purposely delayed, it is important that associated equinus, valgus and cavus deformities of the feet, dislocation of the hips, and other musculo-skeletal deformities be properly splinted and treated orthopedically early rather than waiting until after operation.

(4) *Observation for Hydrocephalus:* Parents can be taught to measure the circumference of the infant's head at home and to record this once a week. They are also instructed in recognition of signs of increased intracranial pressure: bulging of the anterior fontanelle, dilatation of scalp veins, downward displacement of the eyes, vomiting and hyperirritability.

Operation

The primary purposes of operative treatment are removal of the protruding, disfiguring mass and prevention of infection. If this is kept in mind, the importance of tight closure of the sac supplemented by firm approximation of the skin and subcutaneous tissue without tension becomes readily apparent. The operator must constantly be alert also to the danger of actually increasing the neurological deficit by inadvertent damage to intact neural tissue in the performance of an adequate technical repair.

Operations are ordinarily carried out with the patient in the prone position under general anesthesia. We prefer ether administered through an endotracheal tube in all patients except those in the new-born period, where local procaine infiltration plus brandy or paregoric sedation is usually adequate. If the lesion is large or a difficult dissection anticipated, it is well to prepare for constant intravenous fluid administration and possible transfusion during operation.

An elliptical incision is outlined, usually parallel to the longer axis of the base of the lesion. In cervical and thoracic meningoceles this often proves to be in the longitudinal plane, but in the more frequent lumbar and lumbo-sacral lesions (Figure 30A), a transverse incision kept as high as possible above the intergluteal fold is desirable. The skin should be widely prepared and draped, as generous incisions are often indicated to permit wide undermining of skin flaps for closure without tension. Occasionally, rotation or sliding skin flaps, or relaxing incisions may be used to facilitate closure (Figure 32), but in general if such extensive plastic repair is necessary it is better to delay the surgery.

The original incision is carried down through all layers of skin to the fascia of the paraspinal musculature at a point away from the meningocele sac. At this level, dissection is continued until suitable skin flaps have been developed on either side of the lesion (Figure 30B, C). The neck of the sac is then completely freed from adherence to surrounding structures (Figure 30D). Care must be taken during this dissection to avoid injury to any nerve element that may emerge from the sac.

The sac itself is opened, preferably in a longitudinal plane and preferably in the mid-line, but always in an area which appears to be thin and free of neural tissue. It is well to have the head of the operating table tipped downward somewhat to lessen the rapid escape of spinal fluid. The opening into the sac is enlarged carefully until adequate inspection of the inside can be carried out (Figure 31E). If the sac is empty except for spinal fluid, it can be amputated immediately in such a fashion as to preserve an adequate margin at the neck for tight closure. ligature better than a suture.

If the sac contains neural elements, or if the latter run in its wall, par-

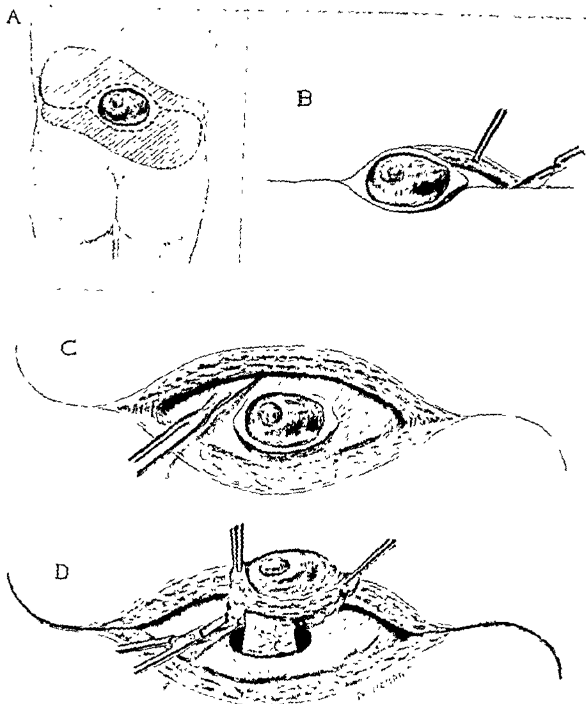


Figure 30 Excision of myelomeningocele — I

A Dotted line indicates skin incision Shaded area shows extent of skin undermining Note ends of *incision curved in opposite directions to increase area of undermining and facilitate* mobilization of skin margins in the center of wound for closure without tension

B Original incision carried down to fascial level

C Skin undermining carried out at fascial level

D Neck of myelomeningocele sac dissected free from attachments to fascia and bony defect *margin*

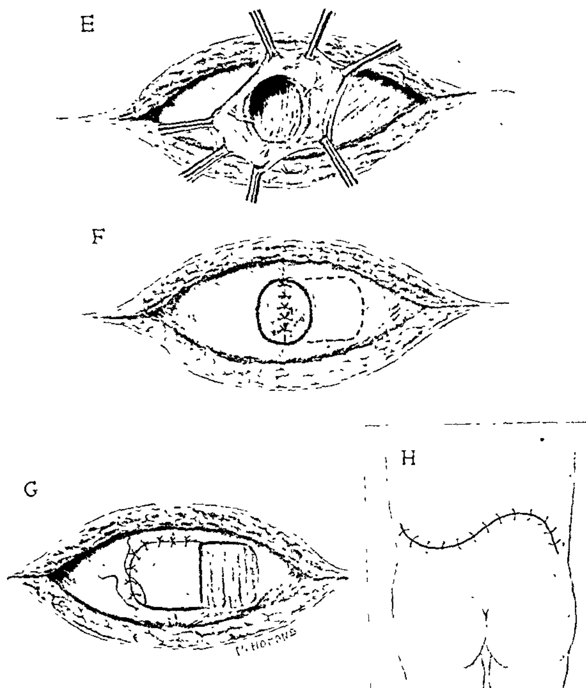


Figure 31. Excision of myelomeningocele — II

E. Sac opened and contents carefully protected. Excess sac and exposed regions have been excised.

F Margins of neck of sac approximated to give water-tight dural closure.

G Dural closure reinforced by fascial flap swung across defect from adjacent paraspinal muscles.

H Two-layer skin closure without tension is most important technical feature of this operation Note diagonal position of wound when this type of incision is closed.



Figure 32A. Two month old infant with large sessile lumbar meningocele. A (*Upper*). Pre-operative appearance.

B (*Lower*). Thirteen days after operation which included V-Y plastic relaxing incision. (A. reprinted by courtesy of The Yorke Publishing Company from *Am. J. Surg*, 75.231-256, 1948.[37])



Figure 33 Lumbar myelomeningocele exposed at operation showing characteristic manner in which roots of the cauda equina may be closely adherent to the wall of the meningocele sac.

ticularly at the neck, special care must be taken. Such neural elements are frequently adherent to the sac but can be carefully mobilized so that they drop freely into the spinal canal. If the neural elements lie actually in the wall of the sac or are inseparable from it, this portion of the sac must be preserved (Figure 33). Faradic stimulation of suspicious structures may be helpful in identifying functional neural tissue. No attempt is made to preserve the entire membranous sac as recommended by some authors, since in our hands this bears no relation to the development of hydrocephalus. As much of the sac is removed in all cases as possible. It is always preferable to leave a little more rather than less of the membranous wall so as to avoid any compression of the neural elements within. Gentle fontanelle or jugular compression by the anesthetist during the repair will demonstrate points of spinal fluid leakage and facilitate water-tight dural closure (Figure 31F).

No attempt is made to repair the bony defect. Under most circumstances no attempt should be made to approximate the paraspinal muscles or their fascia directly across the defect. This is usually impossible and if attempted would compress the space in which the repaired neck of the meningocele sac lies. Where possible it is sound practice to raise a flap of fascia from the paraspinal muscles, leave this attached at one margin of the defect, and suture it across securely to the fascia on the opposite side (Figure 31G). A great deal of attention is then paid to a careful approximation of the skin in two layers. Non-absorbable sutures are used throughout the repair; fine silk or stainless steel wire is used for the skin closure.

Post-operative infection in these wounds comes from without. Its prevention is particularly facilitated by meticulous wound closure to obviate spinal fluid leakage and by careful dressing of the wound to exclude it completely from the perineal region. A strip of vaselined gauze is placed over the suture line, covered with a flat sterile gauze dressing and completely sealed with Elastoplast. A rubber, oiled silk, or plastic sheet is incorporated in the lower margin of the dressing and folded over it in such a way as to waterproof the dressing as thoroughly as possible (Figure 34). If the dressing becomes wet or contaminated at any time it is completely changed immediately.

Post-operative care of these patients is greatly facilitated by placing them face-down on a small Bradford frame supported by blocks on an ordinary crib as shown in the accompanying illustration (Figure 35). The head of the frame should be slightly lower than the foot during the first few post-operative days to minimize spinal fluid pressure at the site of repair. The baby is restrained by cloth binders around the trunk and upper thighs and by well padded wrists and ankle restraints. The baby actually lies on tightly stretched mattress covers protected by a layer of Pliofilm or



Figure 34. Post-operative lumbo-sacral meningocele dressing. The wound is covered with sterile gauze, then Elastoplast. The lower margin of the dressing is sealed from the rectum by water-proof plastic sheet fastened into position with adhesive tape. Diapers are not used in the immediate post-operative period.

other non-irritating plastic film. A suitable gap is left opposite the perineal region so that excreta may fall in a bed pan below. A half-cylinder frame placed over the baby keeps the bed clothes from touching. As soon as the wound is healed and sutures removed, the baby is removed from the frame, and diapers are applied in the usual manner. While on the frame, skin care to the knees, iliac crests and other pressure points is carefully observed. The importance of post-operative wound care cannot be over-emphasized, particularly when large lesions have been removed and closure accomplished only after considerable mobilization of skin. Since the skin is often thin and the deeper layers inadequately closed, skin sutures should ordinarily be left in place at least seven or eight days, we have occasionally used fine wire and allowed the sutures to remain 10 to 14 days. Patients with small lesions repaired easily with no tension do not need to be placed on a frame post-operatively.

The problem of sudden increased intracranial pressure must always be kept in mind during the post-operative period. This, of course, is particularly true in the presence of myelomeningoceles, and will be considered in

more detail in the section on the Arnold-Chiari malformation. It is wise, if the head is larger than normal, if the fontanelle is bulging or the cranial sutures separated at the time of operation, or if ventriculographic studies have shown hydrocephalus whether or not it appears to be arrested, to place these patients on constant ventricular drainage throughout the post-operative period. This can be instituted pre-operatively, at the time of operation, or as soon post-operatively as the fontanelle becomes full. It is carried out by passing a polyethylene catheter into the lateral ventricle through a suture line or drill hole as described elsewhere and attaching this to a closed drainage system (Figure 125). By this means the pressure above the level of the foramen magnum is controlled, as well as the pressure at the site of meningocele repair, at all times. The amount of ventricular drainage daily is measured. If it is small, the drainage bottle is gradually elevated above the level of the ventricles and removed as soon as a 24-hour period with no drainage has been well tolerated. Patients on constant ventricular drainage and all those with extensive lesions are kept on penicillin throughout the post-operative period.

The treatment of spina bifida is not finished with completion of a successful operation. It should be the surgeon's responsibility to see that head measurements are carried out for many months following operation

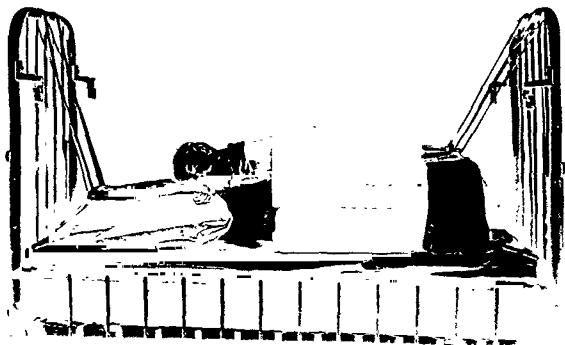


Figure 35. Position of infant post-operatively after removal of lumbar meningocele. Patient is on Bradford frame with legs restrained. No diapers are used. There is a gap in the Bradford frame so that urine and feces may fall into bed pan placed in the proper position. Hood is placed over the frame to support bed clothing. Patient is kept with head slightly downward until the meningocele wound is healed.

to detect the possibility of delayed onset of hydrocephalus. It should also be his responsibility to see that orthopedic help is obtained to institute physiotherapy, corrective braces, and whatever rehabilitative surgical procedures become necessary, and to see that bladder and bowel training are carried out in patients in whom it is indicated.

ARNOLD-CHIARI MALFORMATION

In 1894, Arnold [7] described a caudal tongue-like projection of the cerebellum overlapping and adherent to the upper cervical spinal cord. A year later, Chiari [30] added a description of elongation and kinking of the medulla oblongata associated with this deformity. In 1907, Schwalbe and Gredig [177] gave a detailed anatomical picture of four cases of what they designated the *"Arnold-Chiari Malformation." Since then, numerous clinical* and pathological reports have appeared elaborating the features of





Figure 36 (*Left*). Sagittal section through the cerebellum and brain stem in an extreme case of Arnold-Chiari malformation (Reprinted through the courtesy of The Massachusetts Medical Society from *New England J Med*, 229 108-14, 1943)

Figure 37 (*Right*) Five years and four months old child with large thoraco-lumbar myelomeningocele and arrested hydrocephalus

this hind-brain malformation and its relation to spina bifida.[1, 60, 61, 102, 121, 138, 154, 169]

The essential characteristics consist in downward displacement of the the tonsils of the cerebellum through the foramen magnum, downward displacement and elongation of the medulla oblongata and with it the fourth ventricle, partial or total obliteration of the cisterna magna by firm adhesions of the cerebellum to the medulla and surrounding dura, downward displacement of the upper cervical spinal cord so that the segmental roots appear to course upward toward the foramen magnum to reach their respective dural exits, and a variable degree of kinking of the medulla on the cord (Figure 36). Usually the fourth ventricle opens caudally, directly into the spinal subarachnoid space well below the level of the foramen magnum, and there is frequently choroid plexus extending down to this point or actually protruding from the ventricle. Stenosis of the Aqueduct of Sylvius has also been reported with this malformation as have a variety of bony defects in the region of the basi-occiput and upper cervical spine. Post-mortem studies have revealed a high percentage of craniolacunia and microgyria.[102]

Internal hydrocephalus is probably present to some degree in almost all patients with Arnold-Chiari malformation. This is usually of the so-called communicating type wherein spinal fluid has access from the fourth ventricle to the spinal subarachnoid space but is prevented from passing forward normally through the basilar cisternal system around the brain stem by obliteration of these pathways at the level of the foramen magnum Obstruction may occasionally occur proximal to this point by obliteration of the lumen or outlets of the fourth ventricle or by stenosis of the Aqueduct of Sylvius.

It is probable that in all patients with spina bifida, certainly in all those with a myelomeningocele, who have hydrocephalus at birth or develop it subsequently, that the Arnold-Chiari malformation exists. In many patients with myelomeningocele, the extent of the Arnold-Chiari malformation is apparently such that spinal fluid circulation is impaired to only a minor degree, if at all. Under these circumstances, removal of the myelomeningocele may precipitate a more severe degree of obstruction of the spinal fluid pathways at the level of the foramen magnum and may be followed by either an acute increase in intracranial pressure or by slowly progressive hydrocephalus. This sequence of events has been observed many times and is undoubtedly the usual cause for development of hydrocephalus following surgery for spina bifida rather than any decrease in available absorptive surface resulting from removal of a meningocele sac.

If a patient with a meningocele or myelomeningocele who satisfies the criteria stated elsewhere for surgical repair shows clinical signs of progres-

sing hydrocephalus, it is unwise to attempt excision of the meningocele sac until the hydrocephalus gives evidence of spontaneous arrest or can be arrested by surgical means. If the latter successfully permits normal spinal fluid circulation and absorption, then repair of the myelomeningocele can be carried out secondarily. If the head is enlarged, but not still increasing progressively, in other words, if arrested hydrocephalus exists in the presence of myelomeningocele (Figure 37), it is usually possible to proceed directly to repair of the latter, but with caution. Under such circumstances it is frequently wise to institute constant ventricular drainage at the time the meningocele is removed, particularly if the sac is a large one. The pressure in the ventricular system is kept down until the spinal wound is healed. If a closed system of ventricular drainage is employed (Figure 125), the collecting bottle can then be elevated gradually and when it becomes apparent that it is no longer needed to prevent excessive intracranial pressure, the drainage is discontinued. By this means it is often possible to avoid acute post-operative block at the level of the foramen magnum (Figure 38).



Figure 38A (*Upper*) Two years and eight months old boy with partial paraplegia and arrested hydrocephalus due to large myelomeningocele and Arnold-Chiari malformation.

B (*Lower*). After excision of myelomeningocele patient developed acute hydrocephalus which was successfully relieved by suboccipital craniectomy and upper cervical laminectomy. This might well have been avoided by constant ventricular drainage during and after excision of the myelomeningocele.

Operation

Under general anesthesia administered through an endotracheal tube, the patient is placed on the operating table in the prone position using a cerebellar head rest. Incision is made in the mid-line from the external occipital protuberance to the spine of about C_4. The muscles are separated

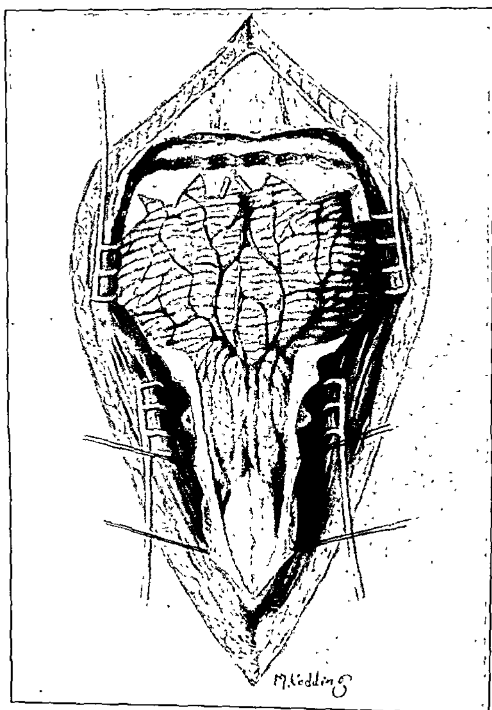


Figure 39. The Arnold-Chiari malformation as exposed by suboccipital craniectomy and upper cervical laminectomy. (Reprinted through the courtesy of The Massachusetts Medical Society from *New England J. Med.*, 229:108-114, 1943)

in the mid-line until the occipital bone and the first three or four cervical vertebral arches are exposed. The atlanto-occipital ligament is usually absent and the thinned-out dura bulges through this space and between the vertebral arches. The occipital bone is penetrated and excised until the dorsal half of the rim of the foramen magnum has been removed and the greater part of both cerebellar hemispheres exposed. The arches of the first, second and third and occasionally the fourth cervical vertebrae are also removed. The arch of C_1 is frequently incompletely ossified. There is usually marked indentation of the dura at the rim of the foramen magnum and at the site of the upper two vertebral arches, producing characteristic waist-like constrictions which persist after removal of the bone.

The dura is incised in the mid-line over the upper cervical cord and the incision carried upward over both cerebellar hemispheres in such a

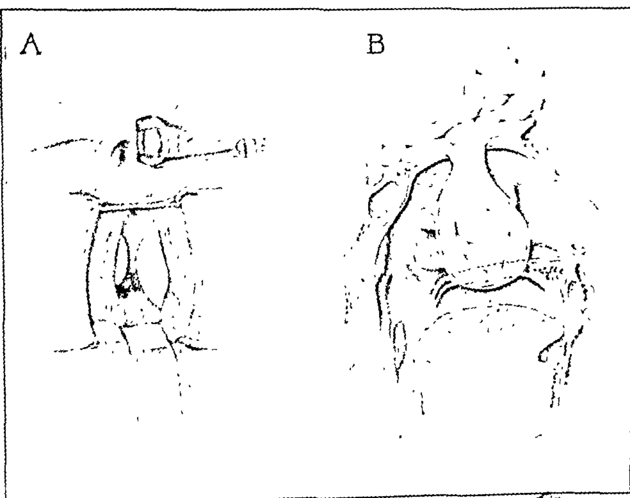


Figure 40A A semidiagrammatic view of encephalocele prot[illegible] the left nos[illegible] through cleft palate.

B A midsagittal view showing the size and position of t[illegible] le in rela[illegible] cribri-form plate and nasopharynx. (Reprinted through the courtesy [illegible] achusetts [illegible] iety from N[illegible] J. Med., 228:8

manner that all areas of dural constriction are released. In young infants, anomalous venous channels in the dura of the posterior fossa may be encountered at this stage and bleed furiously if not properly managed, usually with tantalum clips. Ordinarily, the cisterna magna is completely obliterated. The tonsils of the cerebellum protrude caudally through the foramen magnum to the level of about C_3 and often are tightly adherent to the spinal cord and to the dura (Figure 39): These arachniodal adhesions are removed as thoroughly as possible and the fourth ventricle opened by mobilizing the cerebellar tonsils away from the region of the obex. Meticulous hemostasis is essential to minimize the reformation of adhesions. Amputation of the cerebellar tonsils is ordinarily unnecessary. The dura is left widely open both over the cerebellum and the upper cord. It is wise to suture it back to the muscles on either side. The balance of the wound is closed in layers in the usual manner.



Figure 41. New-born infant with pedunculated meningocele protruding through bony defect at the bregma. This lesion contained no brain tissue and was successfully removed locally. (Reprinted through the courtesy of The Yorke Publishing Company from *Am. J. Surg*, *75:* 231-256, 1948.[77])

In extreme degrees of the Arnold-Chiari malformation, there may also be occlusion of the Aqueduct of Sylvius of sufficient degree to prevent normal egress of spinal fluid from the third ventricle. If this is demonstrated at the time of surgical exploration or by pre-operative ventricular air studies, a Torkildsen type of shunting procedure should be performed at the same time, leading a catheter from a lateral ventricle to the lateral cisternal recess in the cerebello-pontine angle (p. 136).

Occasionally, adequate spinal fluid circulation may not be present in the immediate post-operative period but may establish itself after a period of several days or even weeks. If the deformity includes obliteration of the pontine, lateral and ambient cisternae by displacement or by adhesions, the procedures outlined will, of course, be inadequate. Usually in the latter circumstances, the degree of hydrocephalus, retardation, and neurological deficit of the lower extremities is so extensive that surgical inter-



Figure 42 (*Left*). Pedunculated parieto-occipital encephalocele at birth associated with hydrocephalus due to malformation of the subarachnoid pathways intracranially

Figure 43 (*Right*). Sessile type of occipital encephalocele. This type of lesion is often associated with gross malformation of structures within the posterior fossa. Air studies are desirable prior to removal

vention is undesirable anyhow. If treatment is indicated in such a patient, spinal fluid shunt from the lateral ventricle to the peritoneal cavity is recommended (p. 151).

In general, the results of suboccipital and upper cervical decompression have been disappointing when a severe degree of Arnold-Chiari deformity was found at operation. The patients who have done best have been those with minor degrees of malformation and very slowly progressive hydrocephalus.

CRANIAL MENINGOCELE AND ENCEPHALOMENINGOCELE

All of the mass lesions associated with cranium bifidum have been grouped together in common usage, in our clinic as well as elsewhere, under the term encephalocele. Actually, the different types encountered clinically are analogous to those of the spine discussed in the previous section. More often than not, however, the presence of neural tissue within the lesion is ascertained definitely only when operative or pathological data are available.

Encephaloceles are less common than their spinal counterpart. In this clinic they have been encountered 187 times as compared with 1,157 spinal lesions. They may occur at any point over the vertex or base of the cranium, usually in the mid-line or extending intracranially to the mid-line. By far the most common location is the occipital area where they are apt to be associated with a wide variety of hind-brain malformations. The distribution of our encephalocele cases is seen in the accompanying table (Table VI).

TABLE VI

ENCEPHALOCELE
SITE OF LESION

Level	*No. of Cases*
Nasal	9
Nasopharyngeal	1
Frontal	11
Parietal	27
Occipital	139
Total	187



Figure 44A Huge occipital encephalocele connecting with the subarachnoid space and containing no neural tissues.

Examination

The diagnosis is usually apparent at the time of birth by the appearance and position of the mass. Occasionally, differentiation from a dermoid inclusion cyst (Figure 279) or vascular malformation of the scalp is not possible until x-ray examination or even operation has been performed. Orbital encephaloceles are usually discovered in the investigation of the cause of unilateral exophthalmos. Nasal and nasopharyngeal lesions are apt to be mistaken at first for nasal polyps or some other extracranial tumor obstructing the air-way (Figure 40).



Figure 41B Same patient eight years following removal of lesion No hydrocephalus Development slightly retarded.

Encephaloceles may be pedunculated (Figures 41 and 42) or sessile (Figure 43); they may be completely covered with epithelium only or with full-thickness scalp; they may be partially or totally covered with a transparent, extremely thin, or translucent, parchment-like membrane. Often there are vascular abnormalities of the scalp overlying and adjacent to the sac. The hair may be unusually long and silky immediately about the lesion.

The size of the mass in itself gives no accurate indication of its contents. Huge lesions the size of the baby's head may be entirely filled with fluid (Figure 44) or may have a relatively narrow meningeal stalk (Figure 45), whereas small lesions may contain mostly brain tissue and be associated with gross malformation of vital intracranial structures (Figure 46). It is often obvious on clinical inspection that the sac contains solid tissue, either a large protrusion of brain substance or a thin herniation of cerebral tissue containing part of the ventricular system.

Plain roentgenograms should be made rountinely to determine the size and position of the cranial defect. It is our custom in most encephaloceles, particularly those in the suboccipital region, to perform ven-



Figure 45. Pedunculated occipital encephalocele containing yellow fluid. Atrophic neural and fibrous tissue in stalk have sealed this off from intracranial contents.



Figure 46. Two months old infant with midline occipital encephalocele. This lesion had a narrow neck through the bone and expanded into a much larger meningocele within the posterior fossa which produced marked internal hydrocephalus.

tricular air studies also. A small amount of fluid can be removed from the ventricle and replaced with oxygen or air readily and safely after puncture through a coronal suture. X-ray films are then made in various positions to determine whether the ventricular system is dilated or is distorted in such a way as to indicate direct involvement of brain in the lesion.

Indications for Surgery

Except in instances of obvious gross brain damage where terminal care is imminent, operative treatment is indicated in most patients with encephaloceles. In general, operation can and should be performed earlier in this group of patients than in those with spinal lesions. Operation in the post-natal period should be performed on those lesions with frank or imminent rupture of a thin membrane. Most other encephaloceles can be excised satisfactorily in the first weeks of life, as soon as the baby has stabilized



Figure 47. Antero-posterior and lateral views of three years and two months old child with encephalocele present at the bregma. Large defect in the frontal bone with displacement of the orbital contents laterally on both sides Child of normal intelligence. Essentially normal pneumoencephalogram.

and is gaining steadily in weight. The difficulties of feeding, handling, and ordinary nursing care are such with an encephalocele of any size, and the psychic trauma to the parents is such in the presence of this disfiguring and unconcealable mass, that the earliest possible excision is highly desirable. If it is necessary to wait for some reason, the meningocele sac can be protected in much the same manner as described for spinal lesions.

Many encephaloceles are pedunculated and most of them are at least partially covered with skin. These facts together with the great mobility of the scalp make technical repair after early excision usually a relatively simple problem.

Operation

The general principle of tight closure of the sac supplemented by firm approximation of the skin in two layers enunciated in discussing spinal meningoceles is just as pertinent here. Also it should be reiterated that operation is carried out as prophylaxis against infection and for removal of the mass rather than to improve any neurological deficit which may already be evident. Operations are performed under local procaine or endotracheal ether anesthesia depending upon the age of the patient and

the character of the lesion. If the face-down position is employed, anesthesia is always administered through an endotracheal tube. Wide preparation and draping of the operative field is important.

Elliptical incision in a plane so as to preserve the normal blood supply of the scalp is made. In suboccipital lesions, a longitudinal incision is usually most satisfactory since frequently exploration of the posterior fossa may be necessary at the same operation. Dissection is carried down to the periosteum and then toward the neck of the sac from all sides until the latter is completely mobilized. The sac is then opened and inspected carefully. Neural elements, if present, are freed from adhesion to the sac and replaced intracranially. The sac is amputated leaving sufficient wall at the base to perform water-tight closure. Occasionally a flap of pericranium may be swung across this suture line to reinforce it. No attempt is made to repair the cranial defect at this time in suboccipital lesions.



Figure 48. Encephalocele protruding in the region of the bregma through a skull defect three by four centimeters associated with deformity of the nose. Post-operative appearance after closure of the dural defect and repair of the skull defect by tantalum cranioplasty. Plastic reconstruction of nose to be performed later.

As mentioned previously, encephaloceles in the suboccipital region are frequently associated with hind-brain malformations which may result in obstruction to the circulation of cerebrospinal fluid. It is often wise therefore, particularly if pre-operative air studies have demonstrated a dilated ventricular system, to plan exploration of the posterior fossa at the time of excision of these lesions. By removal of cystic extensions within the posterior fossa, lysis of adhesions of the cerebellum, cisterna magna, and upper cervical cord, division of the tentorium, or other maneuvers, it may be possible to re-establish normal spinal fluid circulation. Partial agenesis of the cerebellum is a frequent finding.

Encephaloceles protruding in the frontal region, at the bregma, into the orbit or through the cribriform plate into the nasopharynx are best

approached through a coronal scalp incision (Figures 47 and 48). The frontal meningocele sac is repaired directly and the scalp replaced. Cranioplasty can then be performed subsequently; both bone grafts and tantalum wire mesh (Figures 49 and 50) have been used successfully in this region. Orbital lesions are approached through a unilateral concealed frontal bone flap (Figure 305) with retraction of the frontal pole extradurally and removal of the roof of the orbit as far as necessary to amputate the lesion and carry out a tight dural closure. Combined intra- and extradural exposure may be necessary. Encephaloceles protruding through the cribriform plate are approached through unilateral or bilateral frontal bone flaps. Abnormal brain tissue is amputated from within and the defect through the cribriform plate repaired by a free or pedicled graft of dura across the defect (Figure 51). These lesions should never be approached transnasally.

There is no particular problem in the post-operative management of most encephalocele excisions. The scalp wounds heal well if they are not under tension. Ventricular pressure should be kept down by intermittent or constant drainage if necessary until the operative wound is healed.



Figure 49A. Lateral roentgenogram of three years and two months old boy with large frontal encephalocele and deformity of the nasal and sphenoid bones

B. Lateral roentgenogram following removal of bone from both temporo-parietal regions with transplantation to the frontal area after excision of the meningocele and repair of the dura. Good cosmetic result. Negative neurological examination.
Same patient as shown in Figure 47.



Figure 50. Antero-posterior roentgenogram of three months old infant after removal of anterior encephalocele, closure of the dura and repair of the bony defect with tantalum mesh. Negative neurological examination.

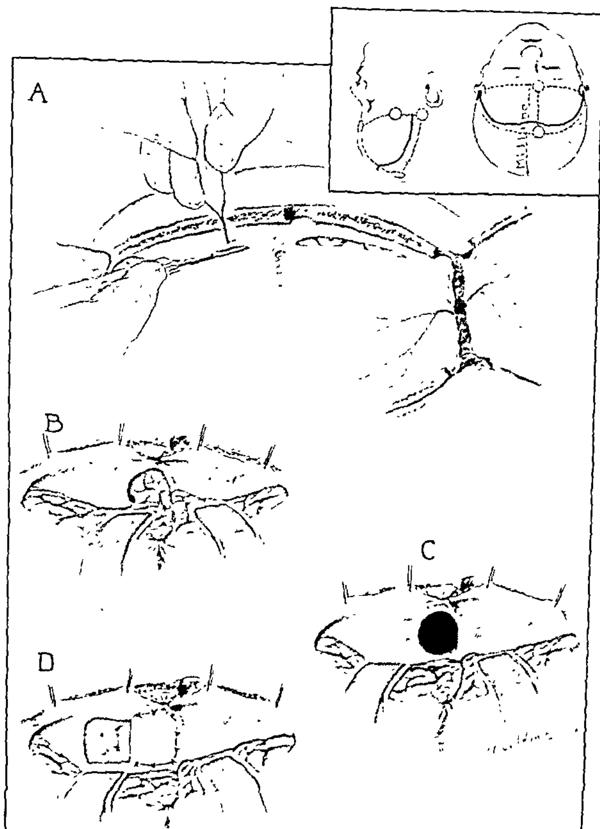


Figure 51 In the insert, the solid line indicates the site of incision, and the dotted lines the limits of the right and left frontal bone flaps In A, the dura has been opened on either side
(*Legend continued on facing page*)

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Chapter II

Diastematomyelia

There is a group of patients with an unusual but consistent type of occult spinal disorder which warrants separate description. The distinguishing feature of this group is the projection of a discrete bony spicule from the posterior surface of a vertebral body through the middle of the spinal canal to divide the meninges into two compartments and transfix the spinal cord or cauda equina. These patients are designated by the term diastematomyelia, which signifies an abnormal cleft or division of the spinal cord or its intraspinal derivatives, in distinction to diplomyelia, which denotes reduplication of the cord.

Diastematomyelia has frequently been described in isolated postmortem examinations of still-born monsters, but has only recently been recognized during life with any frequency as a cause of crippling disability that may be subject to surgical treatment.[87, 131, 136, 137, 160] Twenty-two patients with this congenital anomaly have now been operated upon in this clinic and a number of others have been recognized by roentgen examination.[147] The patients operated upon have been studied and followed in considerable detail and form the basis of this discussion (Table VII). Their age ranges from eight weeks to nine years. There are 17 females and five males. The level of the bony spicule splitting the spinal contents varies from T_5 to L_4 (Figure 52).

The embryology of this particular variety of spina bifida is not clear.[21] Multiple congenital anomalies of the vertebrae, including some degree of incomplete spinal fusion, have been present in every instance. Presumably during the organization of the neural tube from the primitive neuro-ectoderm, aberrant mesodermal cells protrude into the neural tissue on its ventral surface instead of becoming arranged entirely around its periphery. Persisting in this location, they develop into a bony and dural septum.

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of the longitudinal sinus at the anterior limit of the exposure. In B, the pedicle of the herniated brain at its origin from the left frontal pole has been exposed after division of the longitudinal sinus and falx and retraction of the frontal lobes. C shows the inside of the meningocele sac, after amputation and removal of the herniated brain. In D, the dural flap has been reflected from the floor of the frontal fossa across the defect in the cribriform plate and has been sutured to the crista galli. (Reprinted through the courtesy of The Massachusetts Medical Society from *New England J. Med*, 228 815-820, 1943.)

TABLE VII

No.	Name	Age	Level of Lesion	Cutaneous Defect	Neurological Defect	Myelography	Follow-up
1	P. S.	3 3/12	T_{11}	Lipoma	Lower extremities Both sphincters	Not done	Unknown
2.	E. H.	5 4/12	L_4	Tuft of hair	Lower extremities	Not done	7 4/12 yrs. No progression Improvement with tendon transplant
3	G. D.	1 6/12	L_4	Skin dimple	Lower extremities	Diagnostic	5 6/12 yrs. No progression Gait improved
4	M. I.	2 10/12	T_8	None	Lower extremities	Not done	6/12 yrs. No change
5	M. M.	8 weeks	L_4	Tuft of hair and Telangiectasis	None	Diagnostic	3 10/12 yrs. No abnormality Question of muscle imbalance developing
6.	S. S.	2 3/12	L_4	None	None	Diagnostic	2/12 yrs. Continent Asymptomatic
7.	R. A.	1 10/12	T_{10}	Skin dimple	Lower extremities	Diagnostic	3 6/12 yrs. Early improvement subsequently unchanged
8	R. C.	1 9/12	T_{11}	Hemangioma Tuft of hair	Lower extremities Poor sphincter control	Not done	2 5/12 yrs. Improved Only minimal gait disturbance
9.	D. L.	4/12	L_2	Skin dimple Tuft of hair	Urinary incontinence	Diagnostic	6/12 yrs. Improved Bladder probably unchanged
10	J. M.	1 11/12	T_8	Lipoma	Lower extremities	Diagnostic	3 2/12 yrs. Improved
11	A. L.	5 9/12	L_4	Skin dimple Lipoma	Lower extremities Urinary incontinence	Diagnostic	3 2/12 yrs. Improved

TABLE VII–(Continued)

No.	*Name*	*Age*	*Level of Lesion*	*Cutaneous Defect*	*Neurological Defect*	*Myelography*	*Follow-up*
12.	M. R.	$1\frac{1}{12}$	L_3 to L_4	Tuft of hair	Constipation	Diagnostic	$1\frac{8}{12}$ yrs. Definite improvement in gait and normal sphincter control
13.	S. L.	9	L_1 to L_2	None	Lower extremities	Diagnostic	$\frac{1}{12}$ yrs. No change No follow-up beyond 1 month
14.	H. S.	$3\frac{6}{12}$	L_4	None	Lower extremities Incontinent of urine and feces	Diagnostic	$\frac{11}{12}$ yrs. No change
15.	K. H.	$\frac{5}{12}$	L_2 to L_3	Skin dimple	None	Diagnostic	$1\frac{1}{12}$ yrs. No neurological deficit
16.	C. P.	$2\frac{6}{12}$	L_1 to L_2	Tuft of hair	None	Diagnostic	$\frac{1}{12}$ yrs. No follow-up beyond 1 month
17	J. M.	$\frac{4}{12}$	T_5	None	Lower extremities	Diagnostic	1 yr. Improving
18.	K. J.	$2\frac{4}{12}$	L_2 to L_3	Tuft of hair Skin dimple	None	Diagnostic	1 yr. No change
19.	N. T.	$3\frac{4}{12}$	L_1 to L_3	Tuft of hair	Extrophy of bladder	Diagnostic	1 yr. No change
20.	P. M.	$\frac{4}{12}$	L_2	Tuft of hair	None	Not done	$1\frac{1}{12}$ yrs. No change
21.	B. T.	6	L_4	Tuft of hair	Lower extremities	*Diagnostic*	$\frac{8}{12}$ yrs. *Improving* with orthopedic treatment
22.	K. F.	$1\frac{6}{12}$	T_{12}	Lipoma	Lower extremities	Diagnostic	$\frac{2}{12}$ yrs. Improving in gait and bladder control

Diagnosis

As in other types of spina bifida occulta, the symptoms and signs which point to this disorder may be divided into two groups; those associated with a cutaneous deformity overlying the bony anomaly and those associated with abnormal neurological function distal to the level of the lesion.

Cutaneous Defects: These are present in the mid-line or close to it in the same segment as the bony anomaly. The following types of defect have been noted among our cases: abnormal tufts of hair (10 patients) (Figures 53 and 54), dimpling of the skin (six patients) (Figures 54 and 55), subcutaneous, poorly circumscribed fatty tumors (four patients) (Figure 54), and cutaneous blood vessel malformations (two patients) (Figures 54 and 55). The skin lesion may actually be the presenting complaint or it may be noted in the investigation of a neurological disorder.

Neurological Disorders: Most of these patients seek medical care primarily because of impaired function of the lower extremities or of the vesical or rectal sphincters. The commonest complaint is difficulty in walking or learning to walk. Various types of gait disturbance and muscular imbalance in the lower extremities have been noted. Weakness of the anterior tibial and peroneal muscles is frequently found; deep tendon reflexes are occasionally absent in low lumbar lesions and hyperactive in thoracic lesions; atrophy of the lower leg musculature is common (Figure 56); a tendency to drag one leg in walking has been the initial complaint in several patients; numerous types of foot deformity may be seen (Figure 57).

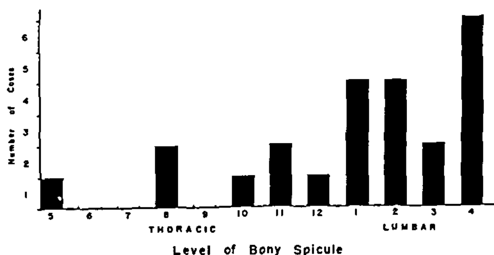


Figure 52. Segmental level of the bony spicule dividing the spinal canal in 22 surgically treated cases of diastematomyelia

In this age group, sensory examination is difficult and unreliable but occasionally may demonstrate clear-cut abnormalities, particularly in the saddle area. Poor sphincter control may be the first abnormality noted in the low lumbar lesions. The diagnosis has been made radiographically several times during the investigation of incontinence.

Lumbar puncture is of little significance in making the diagnosis. The findings are usually entirely normal since there is no obstruction to the circulation of spinal fluid.

X-ray Findings

Pre-operative x-ray diagnosis of diastematomyelia is possible in almost every instance. It is important in the investigation of that the entire spine be examined by x-ray and not just the lumbosacral region. Lateral and oblique films are of little help. However, antero-posterior films usually show very clearly a line of increased density in the center of the spinal



Figure 53. Four year old girl showing abnormal hair growth in the lumbar region associated with spina bidfia occulta and diastematomyelia at L_3.



Figure 54 Five months old infant who shows all of the cutaneous malformations ordinarily seen with diastematomyelia. There is: (1) abnormal growth of hair, (2) a vascular malformation of the skin seen in the midst of the area of abnormal hair, (3) a subcutaneous fat pad just below this area, and (4) a dimple at the upper aspect of the inner gluteal cleft.

canal (Figure 58). Characteristically this is located in an area of spina bifida extending over one or more segments. In addition, there is fusiform widening of the spinal canal which may extend over several segments but is usually maximal at the level of the bony spicule. This widening can be distinguished from that noted in the presence of expanding intraspinal lesions because thinning of the pedicles and erosion of the posterior aspect of the vertebral bodies is not present. In a high percentage of patients, other vertebral anomalies such as incomplete laminal arches, hemivertebrae, fused or small vertebral bodies will also be noted (Figure 58).



Figure 55. Five months old infant with diastematomyelia showing mid-line dimple with "port-wine" stain of the surrounding skin in the lumbar region.

Beautiful visualization of this anomaly may be gained by pantopaque myelography (Figure 59). The opaque medium separates into two columns flowing readily around the mid-line septum. The exact level and extent of the diastematomyelia can be accurately recorded (Figure 60). Myelography is not essential when satisfactory plain x-ray films are available, but is usually a significant aid in planning and performing surgical treatment. It has been performed in 17 of our patients.

Treatment

Normal neurological function of the lower spinal cord and cauda equina depends upon preservation of the differential rate of growth of the spinal column and spinal cord throughout infancy and childhood. It is important, therefore, to relieve if possible any obstruction or distortion to this normal "migration" of the cord which may be producing neurological disorders as long as the patient is still in the period of active growth. The purpose of surgery is prophylactic rather than curative. The patient's parents should understand that operation is undertaken not so much in an attempt to reverse changes already present as to permit subsequent normal



Figure 56. Lower extremities of 4 11/12 year old boy with diastematomyelia. Note the small size of the right lower extremity below the knee. There was a perceptible limp which was the patient's initial complaint.



Figure 57. Deformity of the right foot in 6 8/12 year old girl with diastematomyelia. Note the small size and cavus malformation. Patient had a bony spicule dividing the canal at L_4.

growth and maturation of the nervous system and thus prevent, if possible, increase in loss of function.

Operation is carried out with the patient in the prone position under general anesthesia. An exposure one or two segments above and below the site of the bony spicule is planned. The spicule may or may not be attached to the overlying lamina; more frequently the laminae in this area are defective. Subperiosteal resection of the bony spicule is first performed so that as much of it as possible can be removed before opening the dura.

Intradural exploration should always be carried out. The dura is incised as shown in Figure 61 and the medial reflection adjacent to the bony septum are removed. Numerous adhesions from the cord to these dural reflections are divided. It is then possible to retract the two halves of the cord in turn and continue excision of the bony spicule down to the level of the anterior dura. Following this the two halves of the cord should be freely movable and able to lie in close approximation to one another.



Figure 58 Antero-posterior roentgenogram of the spine in 4 8/12 year old boy with diastematomyelia Note the mid-line bony spicule at T_{11}, widening of the spinal canal from T_{9} to L_{1} without thinning of the pedicles and the double hemivertebra at T_{10}.



Figure 59. Pantopaque myelogram showing division of the column of opaque dye around the mid-line bony spicule and dural septum.



Figure 60 Typical pantopaque myelogram in diastematomyelia. Note division of the column of opaque medium around the bony and dural septum at the level of L_3. The widened spinal canal and spina bifida are also apparent.

No attempt is made to close the anterior dura. The posterior dura is closed in a linear manner, thus converting the two previous channels into a single dural canal. Routine wound closure in layers is performed.

The size of the dural cleft usually approximates that of the bony spicule. However, the division in the cord itself extends for several segments apparently cranial to the bony spicule. In this series of patients, the division has always extended beyond the area of cord exposed. The two halves of the cord always have been joined again immediately caudal to the spicule (Figure 62). In the low lumbar region the septum may actually be through the cauda equina although even here it usually splits a low-lying conus medullaris. In this region, atrophic roots of the cauda equina may be seen adherent to the septum and considerably deflected from their normal courses.

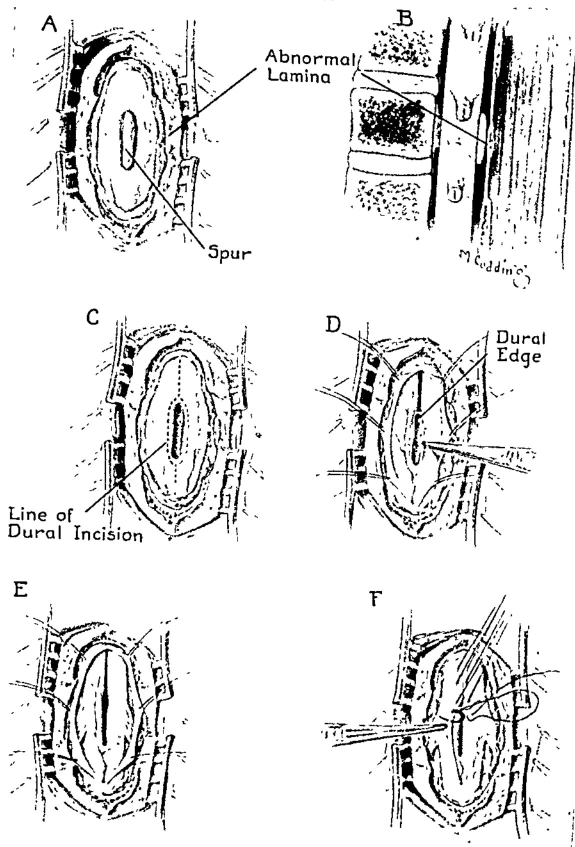


Figure 61. (*See legend on facing page.*)

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None of these patients has exhibited hydrocephalus or other clinical
idence of the Arnold-Chiari malformation.

esults

In the series from this clinic there has been no mortality and no signifi-
nt post-operative complications. Twenty-two patients have been operated
on. One patient was made worse temporarily, but showed subsequent
nprovement. All of the other patients have either improved clinically or

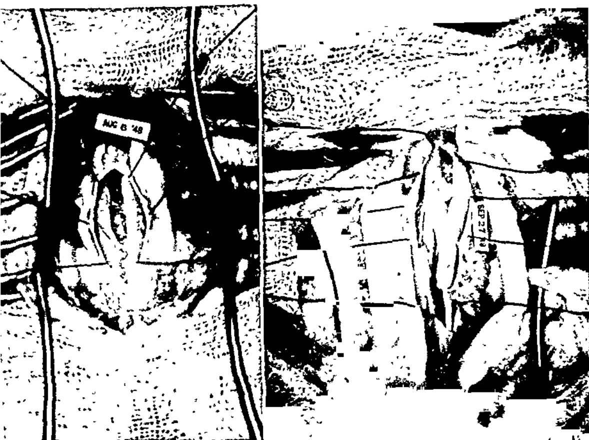


Figure 62. Operative photographs after intradural exposure and removal of the bony and dural septum

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Figure 61A. Bony spur transfixing dura as exposed after laminectomy.

B. Diagram of longitudinal section to show typical relations of bony spur.

C. After extradural removal of most of bony septum, dura is incised as indicated by dotted line.

D Arachnoidal adhesions to medial dural reflections are divided and excision of bony and dural septa completed.

E. Separate halves of cord are now freely movable and approximate one another.

F. No attempt is made to close dura anteriorly; posteriorly it is approximated in linear fashion. Routine wound closure in layers completes operation.

at least have failed to show progression of their neurological abnormalities. The most striking results have been seen in patients with rather rapid increase in bladder and gait disturbance in the weeks before operation. When this loss of function was of recent origin it was almost always reversible. It is emphasized again, however, that the aim of surgery is prophylactic rather than curative so that results should be evaluated in terms of halting or slowing a progressive loss of function.

Chapter III

Congenital Dermal Sinus Tracts

CONGENITAL DERMAL SINUS is the term applied to any depression or tract extending inward from the skin surface which is lined by stratified squamous epithelium. Since the neuro-ectoderm separates from the epithelial ectoderm along the dorsum of the embryo between the third and fifth week of intrauterine life as described elsewhere (p. 5), it is natural to expect that persistent epithelial defects may extend to any depth from the deeper layers of the skin itself down to the substance of the central nervous system, or, indeed, into the central canal of the brain or spinal cord. Also, since mid-line fusion of epithelial ectoderm after cleavage from the invaginated neuro-ectoderm begins in the middle of the embryo and proceeds both cranially and caudally, there is chronologically more opportunity for cutaneous defects to occur in the lumbo-sacral region (posterior neuropore) and next in the suboccipital region (anterior neuropore). This is borne out by clinical observation.

In the sacral region congenital cutaneous abnormalities are extremely common. The great majority are of no neurosurgical significance because they do not extend deeper than the sacral fascia. They are usually lined by hair-bearing skin and may become extremely complex. As such, they are commonly termed pilonidal sinuses and are the source of considerable disability because of their susceptibility to recurrent indolent infections. These superficial dermal or pilonidal sinuses occur much less frequently at other points along the spinal axis as well.

When, however, a persistent defect in the epithelial ectoderm is associated with failure of the mesodermal structures of the same segment to organize and fuse completely, then the dermal sinus may extend all the way into the spinal canal or cranial cavity. Because of its persistent communication with the skin surface, such a sinus tract serves as a constant potential port of entry for infection into the central nervous system and its coverings. In addition to the danger of infection, such a dermal stalk may by its attachment to neural tissue distort the normal growth of the latter.

At any point along a congenital dermal sinus, and particularly at its inner terminus, there may be expansion of the sinus into a cyst. There may be more than one cyst at different levels along the course of the sinus. Such a cyst is defined as an epidermoid cyst if it contains only epithelial

debris and is lined by stratified squamous epithelium alone, or more commonly as a dermoid cyst if it contains in addition to keratin, sebaceous material and hair, and the lining includes hair follicles, sebaceous glands and other elements of the deeper layers of the skin. These cystic expansions may be little larger than the sinus itself or they may be sizeable expanding masses. When such a mass occurs within the cranium or the spinal canal, it may act as any other expanding lesion to interrupt neurological function by local compression of the brain or spinal cord, or it may obstruct the normal circulation of spinal fluid. In addition, it may be the site of abscess formation because of the constant possibility of infection from the skin surface.

Since the first report by Moise in 1926 [142] of an infected sacral sinus which extended through the bone to terminate intradurally and cause meningitis, there have been numerous instances described of dermal sinuses extending into the spinal canal.[110, 124, 173, 199] Most of these patients had some form of infection, meningitis or epidural, subdural or spinal cord abscess, which called attention to the lesion. Others have been discovered and excised during the investigation of spina bifida occulta in patients with neurological disorders, as described elsewhere (p. 11).

Congenital dermal sinuses extending into the cranial cavity are less common. Mount reported two patients in 1949,[145] both of whom had intracranial abscesses and Logue in 1952 collected several more.[126] A number of these patients have been treated in this clinic and form the basis for the following discussion of intracranial dermal sinuses.[135] The subsequent section will deal with comparable intraspinal lesions.

INTRACRANIAL DERMAL SINUSES

The results of treatment of these patients have been so poor when the proper diagnosis was made only after infection had already occurred as compared with prophylactic surgery previous to infection, that the clinical characteristics of this benign but potentially fatal lesion warrant careful description.

Symptoms and Signs

Skin: In or very close to the mid-line, most commonly in the occipital region, a tiny dimple or sinus tract opening is usually seen (Figure 63). Often one or more hairs protrude from the sinus, and there may be an absence of normal hair growth in an area a few millimeters in width adjacent to the opening. It may be impossible to see such a dimple until the area is shaved and examined carefully under good light. There may or may not be "port wine" staining of the surrounding skin. In many patients there



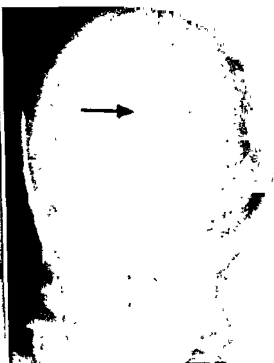


Figure 63 (*Left*). Mid-line skin dimple in occipital region of seven months old infant associated with small subcutaneous dermoid cyst and a tract leading intracranially to a much larger dermoid cyst within the vermis of the cerebellum associated with meningitis and cerebellar abscess formation.

Figure 64 (*Right*). Mid-line, round regular bony defect in occipital region. Usually, defect in bone is much smaller than this and more angulated.

is in addition thickening of the scalp in this region or an actually palpable, small, subcutaneous mass. There may be evidence of superficial infection, such as redness, edema, tenderness or drainage from the sinus opening, or there may have been a history of these findings noted by the family at some time previously. Among 10 patients recently analyzed in this clinic, eight had visible sinus openings, seven of which were in the occipital region and one at the bridge of the nose.



Figure 65. Ventriculogram showing symmetrical dilatation of the lateral and third ventricles with no air entering the posterior fossa in two year old infant with large dermoid cyst within the fourth ventricle.

Intracranial Complications: These include symptoms and signs referable to the presence of an

intracranial mass and those referable to infection; in some patients the clinical picture may be due to a combination of both. Since the mass lesion is usually either a dermoid cyst within the posterior fossa or an abscess associated with a cyst in this area, symptoms and signs may include: headache, vomiting, ataxia, nystagmus, papilledema, extra-ocular motor weakness and enlargement of the head. In other words, the clinical picture is apt to be that of cerebellar or fourth ventricle tumor or simply of obstructive hydrocephalus.

Meningitis may occur, and in several of our patients was the initial complaint. Usually *staph. aureus* or nothing at all is cultured from the spinal fluid. This is in contrast to the meningitis associated with congenital dermal sinuses in the lumbo-sacral area where the infecting organism is usually *b. coli*. In the presence of an intracranial dermoid cyst in a patient who exhibits clinical signs of meningitis, it may be possible occasionally to identify large numbers of epithelial cells in the centrifuged sediment from spinal fluid obtained on lumbar puncture.

Two rather obvious inferences are restated here for emphasis: (1) all infants and children with unexplained meningitis, especially due to *staph. aureus*, should have the entire mid-line of the scalp as well as the skin of the back examined for sinus openings; and (2) in the presence of a mid-line occipital dimple, if there is evidence of increased intracranial pressure it probably means that there is a cystic expansion of the sinus producing obstruction within the posterior fossa.

X-ray Examination

All of these patients have a defect in the skull, obviously, if there is intracranial extension from the skin, but this defect may not always be visible on x-ray examination. The path of the sinus through the bone may be so small and at such an angle that it is obscured in the conventional skull roentgenograms. In young infants, particularly with chronic increased intracranial pressure, the bone may be so thin that a small sinus tract does not stand out by contrast. Usually, however, there is a smoothly outlined, round mid-line defect which varies in size from a barely discernible point of decreased density to a defect one to two centimeters in diameter. These are most apt to be seen in an occipital projection (Figure 64).

Ventriculography which demonstrates symmetrical hydrocephalus of the lateral and third ventricles in the presence of an occipital dermal sinus tract, even if no defect in the bone is visible, means the presence of an obstructing cystic expansion of the tract within the posterior fossa until proved otherwise (Figure 65).

Operation

The treatment of choice is excision of the entire dermal sinus tract, including all cystic expansions wherever they may extend, before infection has occurred, or at least at a time when no infection is present. It is never justifiable to delay exploration of these lesions when there is any possibility of intracranial extension. Therefore, when x-ray films have demonstrated



Figure 66 Extradural dermoid cyst in the posterior fossa of 2 8/12 years old infant connected by narrow stalk through the skull to skin dimple which has been excised with the full thickness ellipse of skin shown. This patient had two previous bouts of meningitis.

a defect in the bone, or when there is any indication of increased intracranial pressure or previous infection, excision of a dermal sinus of the scalp should include preparation to perform primary intracranial exploration as well. For the more common occipital lesions this will involve suboccipital craniectomy for exposure of the posterior fossa.

In the absence of infection, excision of the sinus tract and intracranial cyst is relatively simple. The sinus tract may end blindly in the meninges; it may terminate in an extradural cyst which displaces the dura inward (Figure 66); it may terminate in a cyst within the substance of the cerebellum (Figure 67); it may extend to a cyst within the fourth ventricle (Figure 68). It should be noted that there is often a small dermoid cyst between the scalp and the bone immediately underneath the skin dimple.

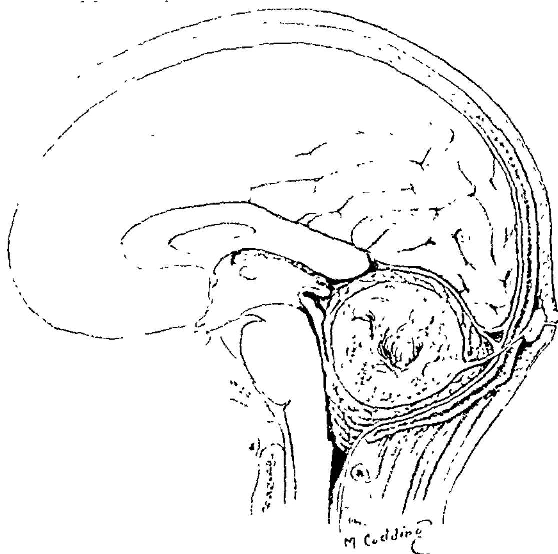


Figure 67. Intracerebellar dermoid cyst causing compression of fourth ventricle and hydrocephalus. Although sinus tract traverses skull in mid-line, cyst may be largely in one cerebellar hemisphere.

It is never safe to conclude that this is the termination of the dermal sinus tract until thorough direct inspection has ruled out any extension through the bone.

If there has been infection, marked inflammatory reaction around the sinus tract (Figure 69) and the capsule of intracranial cysts results and adhesions develop to all surrounding structures. If there has been leakage of the cyst contents into the subarachnoid space, an intense meningeal reaction usually results which may obstruct the normal circulation of spinal fluid (Figure 70). Under these circumstances operation should include lysis of posterior fossa adhesions as thoroughly as possible.

In the presence of gross infection, the contents of the cyst may be liquid and may contain many pus cells as well as bacteria. If such a cyst has not

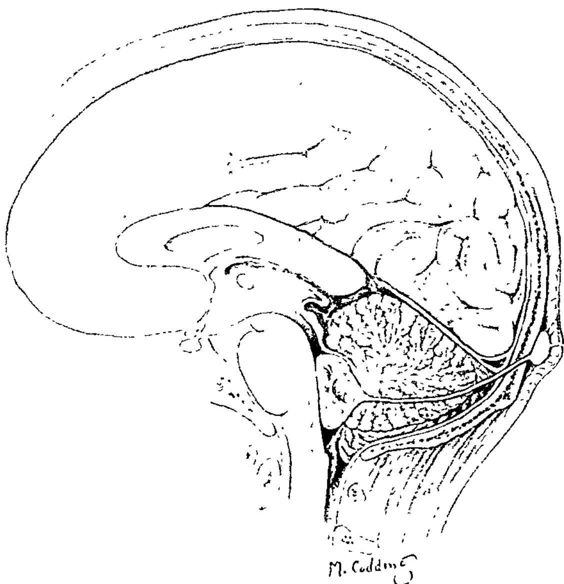


Figure 68 Long sinus tract leading to dermoid cyst within lumen of fourth ventricle causing hydrocephalus.



Figure 69 TBE × 100 Photomicrograph of cross section through dermal sinus extending from skin to a large infected dermoid cyst in the fourth ventricle. Note the tube of squamous epithelium containing desquamated cellular debris There are numerous inflammatory cells in the fibrous tissue surrounding the sinus.

ruptured it may still be possible to remove it completely and, with proper antibiotic protection, this is probably the procedure of choice. If it is necessary to aspirate or drain a cerebellar abscess associated with a dermoid cyst, subsequent removal of the cyst capsule should be performed when the acute process has subsided.

Results

The results of 10 patients with intracranial extension of dermal sinuses treated in this clinic are summarized in Figure 71. It should be noted that in the cases properly diagnosed only after severe infection had occurred,

there were four unsatisfactory results. Two patients died with chronic meningitis and hydrocephalus one month and nine months after operation; a third patient was still alive 18 months after operation but decerebrate as a result of severe meningitis; a fourth patient was cured of meningitis but developed progressive hydrocephalus secondary to virtually complete obliteration of the surface subarachnoid pathways necessitating ventriculo-ureterostomy for relief of increased intracranial pressure. The other six patients are alive and asymptomatic from three months to eight years after surgical excision of the lesion.

INTRASPINAL DERMAL SINUSES

Many of the clinical features of spinal dermal sinuses have already been described in the section on spina bifida occulta (p. 6) since, in the majority of instances, there is a bony defect whenever a dermal sinus tract extends intraspinally. There may occasionally, though, be no more than a groove along one surface of a spinous process, or more rarely, the tract may extend to the meninges through the intervertebral ligaments with no accompanying bony abnormality whatsoever. Intraspinal dermoid cysts with no discernible sinus tract connecting to the skin surface are discussed in the chapter on intraspinal tumors (p. 345).



Figure 70 Base of the brain at autopsy in fatal case. Note obliteration of the basilar cisternae by organized exudate. There was extreme hydrocephalus.

Symptoms and Signs

As previously stated, these sinuses are located predominantly in the lumbo-sacral region, although occasional cases involving the cervical and thoracic regions have been observed (Figures 72 and 73). The sinus opening is in the mid-line or close to it. It is usually not more than a millimeter or two in diameter and may pass unnoticed until local infection occurs. As in the occipital region, there may or may not be thickening, pigmentation, or "port wine" staining of the adjacent skin, and there may or may not be protrusion of hairs from the sinus opening. Frequently the opening is located in the low lumbar region or just at the upper end of the intergluteal cleft (Figure 74).

INTRACRANIAL COMPLICATIONS
OF CONGENITAL DERMAL SINUSES

No	Name	Age	Location of Dimple	Palpable Mass	X-Ray Defect	History of Infection	Location of Intracranial Extension	Result
1	R W	3yrs 1mo	None noted	No	No	None	Cerebellum	Well, 8yrs
2	J P	10mo	Occipit	Yes	No	Meningitis Staph aureus	Cerebellum	Dead, 1mo post op Hydrocephalus
3	A B	2yrs	None noted	No	No	Meningitis Staph aureus	IV Ventricle	Dead, 9mo post-op Hydrocephalus
4	L L	4wks	Occipit	Yes	Yes	Meningitis Staph aureus	Cerebellum and Cisterna Magna	Well, 2 1/2 yrs Has ventriculo-ureterostomy
5	E J	2yrs 8mo	Occipit	Yes	Yes	Meningitis No growth	Post Fossa extra dural	Well, 2yrs
6	J C	1yr 6mo	Occipit	Yes	No	Meningismus No growth	Post Fossa intra dural	Well, 1yr 7mos
7	CDR	6mo	Occipit	Yes	Yes	Abscess Staph aureus	Cerebellum	Alive, 8 mos [illegible]
8	L B	6mo	Occipit	Yes	Yes	None	IV Ventricle	Well, 1yr
9	V K	1yr	Bridge of nose	Yes	Yes	Osteomyelitis No growth	Frontal Fossa extra dural	Well, 9mos
10	W P	8yrs 6mo	Occipit	Yes	Yes	Abscess Staph aureus	Cerebellum	Well, 4mos

Figure 71. Summary of 10 patients with intracranial extension of congenital dermal sinuses.







Figure 72 (*Upper left*). Mid-line skin defect opposite T_1 marking the outer end of a dermal sinus tract leading directly into the spinal cord in a nine months old infant with no neurological abnormality.

Figure 73 (*Upper right*). Eight years and two months old girl admitted because of history of back pain, spasm and fever. Note dimple in mid-thoracic region with cutaneous vascular malformation. Operation revealed dermal sinus tract extending through dura into substance of spinal cord. No cyst.

Figure 74 (*Lower*). Mid-line dimple in lumbar region over area of spina bifida in 1 4/12 year old child. This dimple marked the external opening of a sinus tract which led through the bony defect into the meninges.

There is often a history of redness, swelling and tenderness of the area or of thin or purulent discharge from the sinus opening. There may actually be a history of one or more bouts of meningitis which had apparently been unexplained, or the sinus may first be detected during an acute meningeal infection. The responsible organism is usually *b. coli*, *staph. aureus* or *strep. faecalis*. Often there are signs of meningeal irritation and low-grade fever associated with a spinal fluid pleocytosis but negative cultures.

If the sinus expands intraspinally to form a dermoid cyst (Figure 75), the symptoms and signs may be primarily those of intraspinal tumor; that is, weakness of the lower extremities, pain or paresthesia in radicular distribution, sphincter disturbances, and sensory and reflex abnormalities below a definite segmental level. In the presence of systemic or meningeal signs of infection together with signs of an intraspinal mass in the same patient, an abscess within the cord or within the subdural or epidural space should be suspected and should constitute a real surgical emergency.

X-ray Examination

Spina bifida is visible by ordinary spine roentgenograms in most patients in whom a sinus tract extends in as far as the meninges. In young infants this bony defect may be difficult to see if it is not extensive, particularly in the cervical and sacral regions. Occasionally the sinus opening on

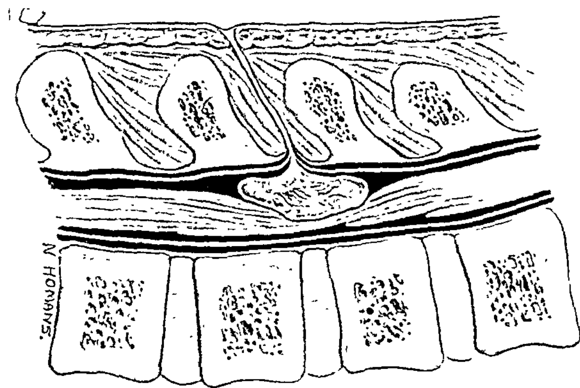


Figure 75. Mid-sagittal section showing expansion of congenital dermal sinus tract to form dermoid cyst in the subdural space at the mid-lumbar level.

the surface may be one or more segments caudal to the level of the spina bifida, since the sinus tract very often courses cranially as it proceeds inward. There may be vertebral anomalies other than spina bifida at the level of intraspinal extension of a cutaneous sinus. If an intraspinal cyst exists, there may also be widening of the canal and thinning of the pedicles.

If plain x-rays are normal, pantopaque myelography may indicate intraspinal extension of a cutaneous sinus by demonstration of a mass or some other type of deformity within the subarachnoid space. Actually, myelography is not often indicated because it is felt that every mid-line cutaneous sinus should be excised to its full extent anyhow.

Operation

As in the cranial lesions, the treatment of choice is complete excision of the entire dermal sinus tract before infection of the central nervous system or its coverings has occurred. Therefore, when mid-line cutaneous defects are associated with an underlying spina bifida with a history of meningitis or superficial infection or drainage, or with symptoms or signs of neurological impairment in the sphincters or lower extremities, or even with fixation of the dimple in the absence of any complication, operation should not be delayed and should include intraspinal exploration. Evidence of progressive loss of neurological function below the level of a cutaneous dermal sinus is indication for urgent operation; transverse myelitis occurring suddenly, especially in the presence of systemic infection, is indication for emergency operation. In the presence of local skin infection or meningitis without neurological abnormality, operation should be deferred until the infection has been treated adequately with appropriate antibiotics and local therapy. *If intraspinal extension is suspected, it is* unnecessary and unwise to probe the sinus tract or inject any type of tracer dye.

Operation should be planned as any formal laminectomy. An elliptical skin incision is made longitudinally to include the skin defect. The sinus tract is traced through the fascia to the region of the spina bifida or to its point of passage through the interspinal ligament. Bony exposure sufficient to trace the tract to the meninges is carred out in standard fashion. The sinus may terminate by expanding into an epidermoid or dermoid cyst extradurally. If the tract is attached to, or enters the dura, it usually does so in the mid-line. The dura should always be opened when this is the case for direct inspection of the intrathecal contents. A longitudinal elliptical incision in the dura is made around the stalk. Often the tract becomes attenuated and ends blindly in the pia, closely attached to the cord. It may seem to penetrate actually to the center of the cord. It may expand intrathecally to form a dermoid cyst either subdurally or within the substance of the spinal cord itself.

Extradural and subdural dermoid cysts, if they have not been grossly infected are usually readily excised. However, if infection has occurred, the capsule of the cyst may be extremely adherent to the meninges, to the surface of the cord, or to the roots of the cauda equina. Under these circumstances it may be necessary to leave parts of the cyst wall behind after evacuating the lumen in order not to risk damage to neural tissues. The same is true to an even greater degree when the cyst is within the cord. Here the cyst should be completely emptied but the capsule removed only in so far as it comes away freely. Attempts to separate the cyst wall from the cord by sharp dissection will almost invariably result in neurological damage.[92] The dura should usually be closed tightly and all communication between the meninges and the skin surface completely excised. Operations for congenital dermal sinuses, whether performed during a period of acute *infection or not, should have the protection of appropriate antibiotic* therapy.

Chapter IV

Craniosynostosis

THE CRANIAL BONES are normally separated from one another at the time of birth. A firm, fibrous union between them usually occurs by the fifth or sixth month of life, but solid bony union is not completed until the sixth to eighth decade. When one or more of the cranial sutures becomes prematurely obliterated, particularly before birth, deformity of the cranial vault results. Such deformities may become extremely grotesque, but of more importance is the effect of this bony anomaly on the growth and function of the central nervous system.

During the first year of life the brain increases about 135 per cent in weight (Figure 76), and during the same interval more than 50 per cent of all post-natal increase in the circumference of the skull occurs.[34, 65] Normal expansion of the skull is directly dependent on normal growth of the brain, and conversely, normal growth of the brain is possible only when the bones of the skull remain ununited and capable of expansion. If premature closure of cranial sutures is present at birth, therefore, or increases in degree during the first year or two of life, significant restriction of the growth of the brain may follow.

Previous to the general use of x-ray examination, a complex descriptive terminology existed which was responsible for much confusion in understanding this disease. Many of these classical descriptive names have persisted in common usage. However, it seems wisest to consider together all the various types of premature closure of the cranial sutures under the term craniosynostosis. Classification is then simplified by grouping cases according to which of the sutures is involved.

It is important at once to differentiate primary craniosynostosis, which results in secondary compression of the brain, from microcephaly, in which early closure of the fontanelles and approximation of the sutures may occur due to failure of the brain itself to grow and push the skull ahead of it. In craniosynostosis, there is ordinarily no involvement of the central nervous system other than restriction of its development by the prematurely fused skull. In microcephaly, there is primary agenesis of the brain with resultant failure of the skull to grow normally (Figure 77). Because of the vast difference in treatment and prognosis, this fundamental difference must be clearly appreciated.

Etiology

The cause of craniosynostosis is unknown. It is due presumably to an inherent mesenchymal defect rather than any known pre- or post-natal disease process. Since 1851 when Virchow [195] suggested that it might be associated with meningitis, it has been variously attributed to syphilis, rickets, pre-natal or birth trauma, endocrine dysfunction and intra-uterine infection. However, none of these has been substantiated. Repeated studies of the bones involved have failed to reveal any gross or microscopic abnormalities of significance.[78]

The probability that this lesion is actually the result of some fundamental defect in germ plasm is supported by its frequent occurrence in siblings and successive generations, by its greater incidence in males (85

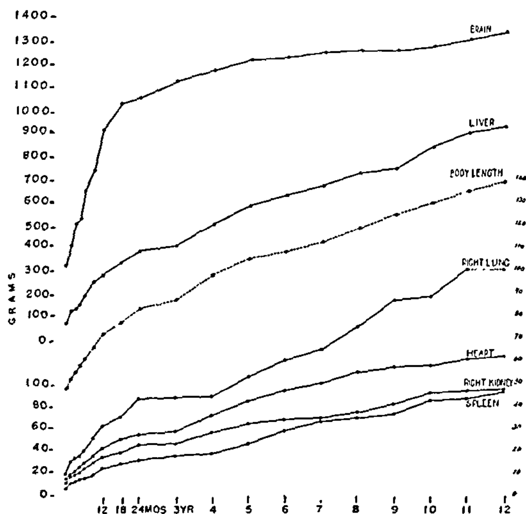


Figure 76. Chart showing rapid increase in brain weight in first two years of life compared to other organs of the body Growth of the brain continues but at a much slower rate from two to 12 years of age. (After Coppoletta and Wolbach, *Am J Path*, 9 55-70, 1933.[a])



Figure 77. Antero-posterior and lateral roentgenograms of one year old child with microcephaly. Note symmetrical small calvarium, sutures are all open but less prominent than normal. Grossly retarded development.

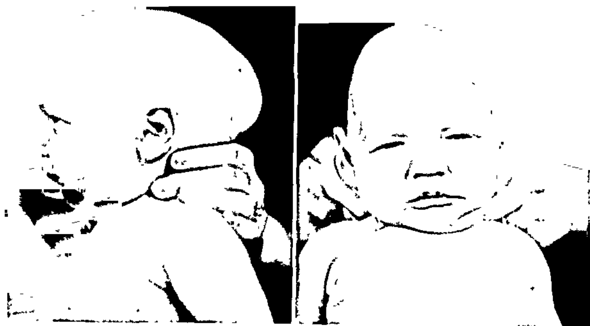


Figure 78. Lateral and anterior views of one month old infant with premature closure of the sagittal suture. Note marked elongation of antero-posterior diameter and narrowing of the biparietal diameter of the head.

per cent) than females, and by the common association of other congenital anomalies, particularly syndactylism.

Symptoms and Signs

The important clinical characteristics of this anomaly are related to: cranial deformity, eye abnormalities, mental retardation, convulsions, and the presence of associated congenital lesions.

Cranial Deformity: The severity of the deformity depends upon the age at which premature fusion begins. The deformity is extreme only when craniosynostosis is present at birth. Distortion present at birth becomes much more accentuated during the rapid growth of the first year of life if no treatment is carried out.

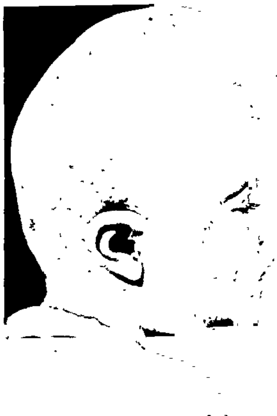


Figure 79. Characteristic appearance in a 1 11/12 year old child of severe deformity due to premature closure of the coronal suture. There is marked brachycephalic deformity of the head with increase in the biparietal diameter (Reprinted through the courtesy of The C V. Mosby Company from *Surgery*, 24.518-541, 1948)

The type of cranial deformity depends upon which sutures are involved. Virchow recognized that when one of the cranial sutures closed before the rapid growth of the brain was completed, the skull expanded in a direction axial to the closed suture; that is, if growth was restricted in any one direction, an attempt at compensation resulted in overgrowth in whatever direction expansion was still possible. Thus, when the sagittal suture closes permanently, lateral growth is limited and unusual antero-posterior expansion occurs. The skull appears elongated and narrow, or dolichocephalic (scaphocephaly) (Figure 78). If the coronal suture fuses prematurely, antero-posterior growth is limited, and there is unusual lateral expansion so that the skull appears wide or brachycephalic with a broad flattened forehead (acrocephaly) (Figure 79). If there is fusion of the coronal and perhaps also the squamosal suture on one side only, a striking and characteristic asymmetry of the skull results with flattening of one side of the forehead (plagiocephaly) (Figure 80). When there is fusion of all the cranial sutures, there



Figure 80. Two months old infant with premature fusion of the right coronal suture only. Note flattening of the right forehead, prominence of the lower right temporal region and asymmetry of the face and orbits.



Figure 81. Two years and seven months old child with premature fusion of all the cranial sutures and deformity of the sphenoid and orbital bones. In addition to the exophthalmos there was low grade chronic papilledema.

is restriction of growth in all directions, and the skull expands in the direction of least resistance – upwards against the region of the anterior fontanelle. The skull becomes pointed or tower shaped, the base is pushed downward, the sinuses remain undeveloped and the orbits shallow (oxycephaly) (Figure 81).

Cranial deformity may usually be noted shortly after birth. Often this deformity is considered due to molding of the head during delivery. Certainly when abnormal shape of the head does not disappear during the early post-natal period, x-ray examination should be performed without delay to rule out craniosynostosis.

Eye Abnormalities: Because of the prominence of ocular complaints many patients with craniosynostosis are first seen by the ophthalmologist. Eye abnormalities include exophthalmos, papilledema, optic atrophy and disturbance of extra-ocular movements. Exophthalmos is due to the extreme shallowness of the orbits which results from pressure downward on the supra-orbital plates, and deformity of the orbital bones themselves. This often displaces the eye-balls laterally as well, thus producing a divergent strabismus and sometimes limitation of eye movements. Papilledema occurs occasionally in patients with fusion of all cranial sutures who develop intracranial pressure as growth proceeds. This papilledema is usually of a chronic variety without hemorrhages. Optic atrophy accompanied by loss of visual acuity results eventually in patients with neglected increased pressure from craniosynostosis (Figure 82). Complete blindness has been known to ensue before the correct diagnosis was made.

Mental Retardation: There is a high incidence of mental deficiency among patients with craniosynostosis if surgical treatment is not performed in early life. The extent of mental retardation is probably roughly proportional to the degree of restriction of the normal rate of brain growth. Thus, it is most apt to be marked when premature fusion of several sutures is present before birth. It has not occurred when only one coronal suture was prematurely fused. Time after time in this clinic it has been noted that the normal processes of development and learning have seemed to accelerate immediately and dramatically after craniectomies have been performed, not only in patients with multiple suture involvement but even for simple fusion of the sagittal suture alone or bilateral coronal sutures alone.

Convulsions: Seizures are not common in craniosynostosis and have been seen ordinarily only in chronic neglected cases where extensive cortical atrophy has occurred.

Associated Anomalies: Congenital anomalies elsewhere in the body unquestionably occur with greater frequency in patients with craniosynostosis than they do in the general population. For some reason which is not clear

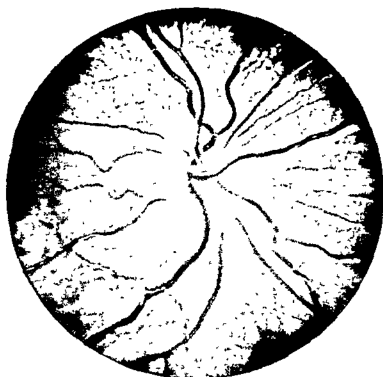


Figure 82. Photograph of the fundus of a five year old child with severe chronic papilledema and optic atrophy associated with craniosynotosis involving all the cranial sutures.

this is particularly true when the coronal suture alone is involved. Syndactylism of the hands or feet is extremely common; when it occurs, it is usually symmetrical. Premature fusion and distortion of the bones of the face also occurs frequently when the coronal or all of the sutures are fused; it never seems to occur when the sagittal suture alone is involved. Various types of deformity of the facial bones are seen. Usually there is overgrowth of the lesser wing of the sphenoid, depression of the nasal and malar bones, with a high arched palate and choanal atresia. The latter deformity may cause marked obstruction to the nasopharyngeal airway and a persistent mucoid nasal discharge. Other anomalies seen in this clinic associated with craniosynostosis include cleft lip, cleft palate, spina bifida, congenital heart disease, and genital abnormalities.

X-ray Findings

The diagnosis of premature fusion of the cranial sutures can always be made and the type determined by x-ray examination. In young infants particularly, x-ray study should be used if there is any question of structural deformity of the skull. Roentgenograms confirm readily the contour of the skull, which may be indefinite on clinical examination because of hair, facial abnormalities, edema of the scalp, or other reasons. In ad-



Figure 83. Antero-posterior and lateral roentgenograms of two months old infant with premature fusion of the sagittal suture alone. Note characteristic shape of the skull and increased density adjacent to solidly fused sagittal suture.

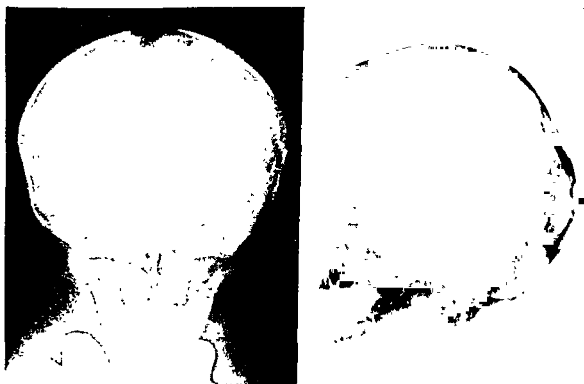


Figure 84. Antero-posterior and lateral roentgenograms of 10 months old child with premature closure of the coronal suture, bilaterally. Note characteristic shape of the skull, foreshortened frontal fossa, deflection of the sphenoid wing and small size of the orbits. (Reprinted through the courtesy of The C V. Mosby Company from *Surgery*, 24 518-541, 1948.)



Figure 85. Antero-posterior and lateral roentgenograms of infant with premature fusion of left coronal and squamosal sutures. Note marked asymmetry of skull with small frontal and large temporal fossa on the left.

dition, satisfactory films demonstrate which sutures are patent and which have closed prematurely (Figures 83, 84, 85, 86 and 87). In the region of a fused suture there is increased density, and often actual mounding up of the bone either externally or internally. Premature obliteration of the anterior fontanelle is also readily detected.

If there has been restriction of brain growth with increased intracranial pressure, exaggerated digital markings will be visible over the convexity as well as thinning of the posterior clinoid processes. Skull roentgenograms also demonstrate frequently associated abnormalities of the orbital, nasal, sphenoid, and maxillary bones. Choanal atresia can be confirmed by transnasal lipiodol injection. Ventricular air studies are ordinarily not performed, but may be of some prognostic significance in chronic cases with mental retardation.

Treatment

A wide variety of surgical procedures has been employed since 1890 [117, 118] in an attempt to relieve the cerebral compression and cranial deformity resulting from premature fusion of the sutures. Subtemporal decompression on one or both sides, elevation of large bone flaps on one or both sides,[56] circular craniectomy, removal of several plaques of bone, creation of artificial sutures,[65, 66, 90, 117, 144, 181] and "morcellation" of the entire calvarium [111, 112, 204] have all been used. In the presence of increased

intracranial pressure, particularly in older children, these procedures have all met with a certain amount of success. However, in the first year of life most of them have been unsatisfactory because of the extremely rapid regrowth of bone which obliterates the artificial defects.

In early life new bone grows very quickly from the outer layers of dura as well as from periosteum. Firm bridging of operative defects may occur *within three months, and solid bony union across craniectomies* usually occurs within six months, thus preventing any further skull expansion. This necessitates repeated operations if growth is to continue.

Because of the great desirability of performing craniectomies within the first few months of life in order to achieve optimum results, efforts were made in this clinic to devise a means of preventing or retarding significantly the rapid regeneration of bone characteristic of this age group. It was found in the laboratory that three inert foreign materials, tantalum, Leucite, and polyethylene placed over the margins of artificial sutures in animals would retard their union.[97] Polyethylene in the form of a thin *film was found most satisfactory for technical reasons, and has been used* routinely since 1947.[90]

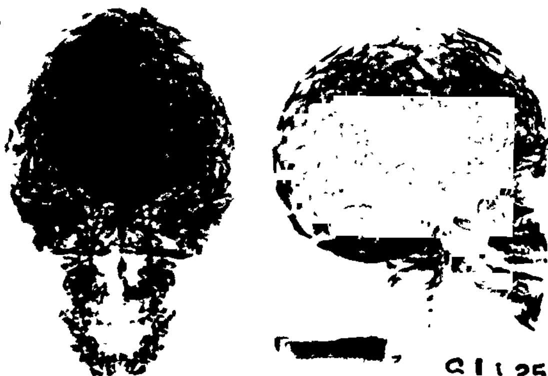


Figure 86. Antero-posterior and lateral roentgenograms of 6 3/12 year old boy with neglected premature fusion of all the cranial sutures This boy was markedly retarded and almost blind X-rays shows small head with extreme diploic markings, marked *impression of the venous* sinuses in the skull and no evidence of any of the cranial sutures (Reprinted through the courtesy of The C. V. Mosby Company from *Surgery*, 24.518-541, 1948)

The general plan of treatment is to perform a linear craniectomy in the region of the prematurely fused suture. This is accomplished by making several burr holes, three to five centimeters apart. A strip of bone, one to one and a half centimeters in width is then removed by cutting between the burr holes either with stout scissors or a Gigli saw. Pericranium is removed as widely as possible and always for a distance of at least two to three centimeters back from the margin of the craniectomy. Drill or punch holes are placed at intervals along either margin of the craniectomy.



Figure 87. Antero-posterior and lateral roentgenograms of two weeks old infant with premature fusion of coronal and sagittal sutures together with marked deformity of the sphenoid bone.



Figure 88. Method of preparing polyethylene film for use in craniosynostosis. The film is placed over the edge of a malleable abdominal retractor and held in this position with string. After boiling for 20 minutes, the film maintains its shape when removed from the retractor, as shown in the foreground.

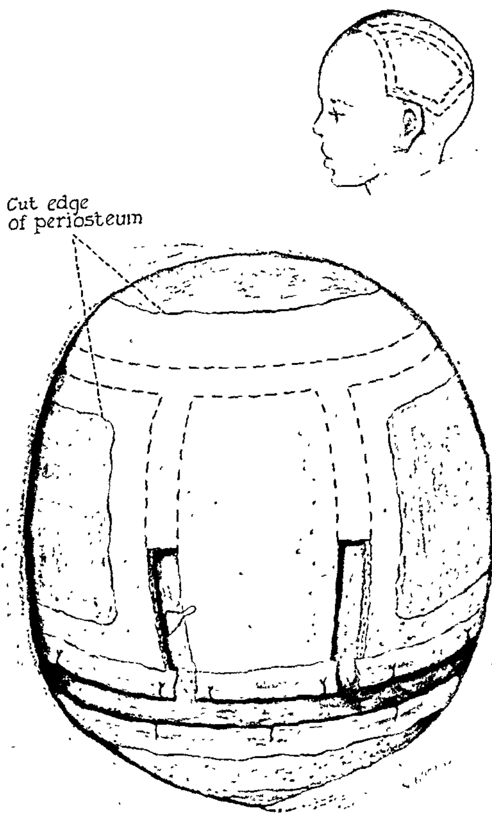


Figure 91. *(See legend on facing page.)*

The polyethylene film is prepared as follows. It is cut into strips two and a half centimeters wide and 15 to 25 centimeters in length. Each strip is folded over the edge of a malleable abdominal retractor bent according to the contour of the infant's head (Figure 88). This is wrapped in several layers of gauze and placed in boiling water for 30 minutes. The sterilized film then maintains its shape when it is removed from the malleable retractor and is ready to be slipped over each margin of the craniectomy. If handled carefully, the film lies smoothly over inner and outer bone surfaces and is held in position by silk sutures passed through the punch holes and film. The film is placed over both edges of each craniectomy. The dura, of course, is never opened.

Thorough hemostasis of these large exposures should be accomplished before wound closure. Even then, it is usually necessary to aspirate fluid collections under the scalp on two or three occasions during the postoperative period. Care should be taken not to apply head dressings too tightly as the scalp may be stretched perceptibly after this operation and additional pressure from the outside can easily cause scalp necrosis. The unavoidable loss of blood during these operations, in young infants particularly, is apt to be greater than the surgeon suspects, so that whole blood transfusions should always be given.

Sagittal Suture: The patient is placed on the operating table face down with the head slightly extended. A mid-line incision is made from the hair-line back to a point about three centimeters posterior to the lambdoid suture. With scalp flaps widely reflected, the pericranium of the entire vertex is stripped off and bleeding controlled with bone wax. Linear craniectomies are made in the manner previously described about two centimeters to either side of the mid-line (Figure 89). It has been felt wise to leave a strip of bone over the longitudinal sinus. It is most important to be sure that the craniectomies extend well anterior to the coronal suture and posterior to the lambdoid suture.

Coronal Suture: With the patient in the supine position a coronal incision is made back of the hair-line. Pericranium is widely excised and linear craniectomy performed in the region of the fused coronal suture. This should extend well beyond the inferiorly displaced squamosal suture in each temporal region (Figure 90).

Sagittal and Coronal Sutures: Operation is usually performed in two stages, particularly in very young infants. In the latter it can all be done

←

Figure 91. Operative field in premature closure of all the cranial sutures. The first step has been completed including lambdoid craniectomy and half of the parasagittal craniectomies. At the second stage, coronal craniectomy and the balance of the parasagittal craniectomies as shown in the outline are completed. Polyethylene film is used to line all of the bone margins. Insert shows position of the coronal, lambdoid, squamosal and parasagittal craniectomies at completion of second stage.

through a coronal incision from ear to ear. The lambdoid craniectomy and *posterior half of the parasagittal craniectomies are* performed at one sitting and the coronal and balance of the parasagittals a week later (Figure 91). In older children separate scalp incisions may be necessary. If the squamosal sutures are fused they can be opened and subtemporal decompressions also done at this time.

Orbital Decompression: Associated particularly with fusion of the coronal suture there is often overgrowth of the lesser wing of the sphenoid, downward displacement of the orbital roof and consequent exophthalmos due to the very small orbits. In these circumstances it is felt that orbital decompression should be performed in addition to coronal craniectomy as early in life as possible. In young infants this is planned as a two-stage operation. A coronal incision is made with the baby in the supine position. Coronal craniectomy is first performed on one side. The temporal muscle is reflected and a subtemporal decompression carried out. The hypertrophied, keel-like projection inward of the lesser wing of the sphenoid is then removed together with the bony roof of the orbit. The margins of the coronal craniectomy are covered with polyethylene film in the usual manner and the wound closed. *A week later, the coronal incision is reopened* and the same procedure carried out on the opposite side. This type of two-stage procedure has been well tolerated within the first month or two of life, but it is emphasized again that it should not be attempted without the support of whole blood transfusions.

Results

In this clinic, 120 patients with craniosynostosis have been treated by surgical procedures. The age distribution is seen in Figure 92. The propor-

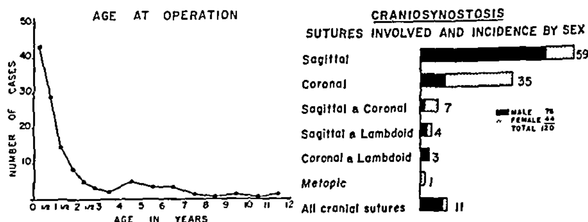


Figure 92 (*Left*). Age at the time of operation in 120 children with premature fusion of the cranial sutures

Figure 93 (*Right*). Incidence of the various sutures involved in 120 children treated surgically for craniosynostosis The tendency of the coronal suture to be involved more in females is of interest

tion of the various types of craniosynostosis is noted in Figure 93. Before 1947 a number of different types of operation were performed and many of them were on youngsters varying between three and seven years of age. Since 1947, linear craniectomy with insertion of polyethylene film has been performed on all patients treated, a total of 84. Also during this period the age at time of operation has been much less. A higher and higher percentage of patients is being seen and operated upon under the age of one year. Thirty-five have now been operated on in the first six months of life.



Figure 94. Severe deformity due to premature closure of the sagittal and coronal sutures at birth.

A (*Upper*). Pre-operative and two-week post-operative clinical appearance.

B (*Lower*). Pre-operative and post-operative x-ray appearance. Note immediate elevation of the vertex and resumption of more normal skull shape.

There has been no operative mortality. Post-operative complications have been few. Many of the scalp flaps require post-operative aspiration on several occasions because of the accumulation of serosanguinous fluid. There have been two post-operative wound infections requiring removal of the plastic film and another infected subgaleal fluid collection which subsided without the film being removed.

Since the use of polyethylene film together with wide excision of periosteum, a second operation has been necessary in only a few instances. If the initial operation is performed during the first two to three months of life, it is likely, even with the film, that reopening of bridged craniectomies may be necessary before two to three years of age. However, ordinarily the craniectomies will remain open until the important period of rapid skull expansion has been passed.

The prognosis for normal mental development of a patient with cranio-*synostosis treated during the first year of life is excellent.* After this period the percentage of patients with retardation increases markedly. It has been surprising, however, to note the acceleration in development and also improvement in social behavior which appears to occur after craniectomy, even in the older age group.

The cosmetic results of operation vary directly with the age at which surgery is performed. The most striking improvement in skull contours and exophthalmos is obtained when patients are operated upon during the first one to three months of life (Figure 94). If operated upon after the age of two years, little change in the shape of the skull may be expected. The pre- and post-operative appearance of representative patients of the various types of craniosynostosis are shown in Figures 95, 96, 97 and 98.



Figure 95. Premature closure of sagittal suture.

A (*Upper*). Appearance at five months of age.
B (*Lower*). Appearance five years after bilateral parasagittal craniectomy with insertion of polyethylene film.

(A. reprinted through the courtesy of The C. V. Mosby Company from *Surgery*, 24:518-541, 1948.)

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(A. reprinted through the courtesy of The C. V. Mosby Company from *Surgery*, 24:518-541, 1948.)









Figure 96A (*Upper*). Appearance of 10 months old infant with closure of coronal suture but no involvement of facial or orbital bones Coronal craniectomy performed.

B (*Lower*). Appearance at two years of age. Head has grown satisfactorily and mental development appears to be close to normal



Figure 97. Unilateral coronal synostosis on the left.

A (*Upper*). Pre-operative appearance.

B (*Lower*). Appearance 18 months after left coronal craniectomy and subtemporal decompression.



Figure 98A (*Upper*) Three months old infant with premature craniosynostosis involving the coronal suture and both sphenoid bones The metopic suture remained widely patent. There *was exophthalmos, choanal atresia and bilateral syndactylism of the hands and feet.*

B (*Lower*). At three and a half years of age Treated by coronal craniectomy, transfer of both frontal bones to meet in the mid-line, and bilateral orbital decompression at three and a half months of age

Chapter V

Congenital Defects of the Scalp and Skull

CONGENITAL ABSENCE of areas of the scalp may occur independent of other abnormalities. Such defects are usually small and usually in the parietal region. They may or may not be in the mid-line. They are of little clinical importance if the underlying structures are intact but they should always alert the observer to associated anomalies. These scalp defects are usually filled by a parchment-like membrane and there is apt to be abnormal vascularity or hair growth in the surrounding skin (Figure 99). Treatment consists in simple excision and repair of the scalp defect when clinical and x-ray

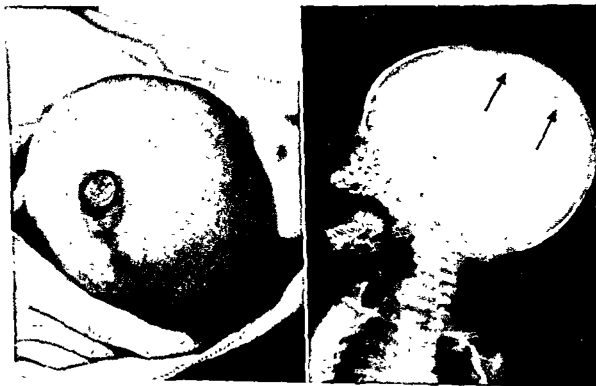


Figure 99 (*Left*). Parchment-like area of complete absence of skin in the right parietal region. No defect in the underlying bone.

Figure 100 (*Right*). Lateral roentgenogram of seven months old infant showing congenital absence of a large portion of the parietal bones. Size and shape of the cranial vault are otherwise normal.

studies and direct operative observations have ruled out underlying lesions.[107]

Congenital absence of a portion of the skull may occur with no associated abnormality of the brain, meninges or scalp. Such defects have been seen in the mid-line varying from a centimeter up to 8 to 10 cms. in diameter (Figure 100). This probably represents a simple fusion defect comparable to spina bifida occulta.

Persistent skull defects are also seen occasionally in the region of the parietal emissary vein foramina. They are usually bilaterally symmetrical (Figure 101). These defects decrease in size somewhat as growth proceeds but do not close completely. Unless they are unusually large, they are of



Figure 101 Antero-posterior roentgenogram of the skull of a two year old infant with persistent bilateral parietal foramina. Patient asymptomatic and normally developed.



Figure 102A (*Left*). Lateral pneumoencephalogram of 1 7/12 year old boy born with a large congenital defect of the parietal bones bilaterally. Normal development and negative neurological examination.

B (*Right*). Lateral roentgenogram of same patient, now nearly six years of age, following tantalum cranioplasty.

no clinical significance and no treatment is necessary. Similar persistent defects in the region of the mastoid emissary foramina are much rarer.

It is characteristic of congenital skull defects that spontaneous obliteration as a result of normal bone growth does not occur. Whereas a traumatic or surgical defect of the cranial vault in an infant fills in rapidly with new bone arising from the dura and from the margins of intact skull, this does not take place in congenital lesions. Therefore, if these defects are of appreciable size, cranioplasty by one of the methods described elsewhere (p. 173) is indicated. In general, it is desirable to wait until a child is three to four years of age and the skull has passed through its period of rapid expansion before cranioplasty of congenital defects is undertaken (Figure 102). External protection can be used during this interval if the defect is a large one.

When congenital absence of a portion of the skull is accompanied by absence of the scalp as well, treatment is more complicated. If the dura is intact, the problem is primarily prevention of infection and subsequent plastic repair. Surprisingly large scalp defects, if kept clean and free of devitalized soft tissues, will granulate and epithelialize readily, permitting cranioplasty to be performed later if necessary. Occasionally, in defects at





Figure 103 (*Upper*). Two weeks old infant born with congenital absence of the scalp and cranium and defective meninges over the vertex. Thick eschar has formed due to drying of secretions in the defect. Severe hemorrhage occurred on three occasions due to rupture of vessels at the posterior edge of the defect in the mid-line.

Figure 104 (*Lower*). Congenital defect of the skull and scalp. View of infant at seven months of age. The defect is now entirely covered with epithelium but there is no bone over the vertex. Cranioplasty to be performed later. Same patient shown in Figure 103.

the vertex, it may be wise to do a Z-plasty of the scalp or swing a pedicle flap to cover the defect more quickly.[107] In congenital scalp defects where the underlying bone is deficient and subsequent cranioplasty may be necessary, it is wise to avoid split-thickness grafts.

A much more critical lesion for the patient is the occasional absence of scalp, bone and dura over the vertex. This may occur with no developmental defect of the brain itself, but unless managed carefully it is apt to be a fatal lesion. The exposed area of brain is of course vulnerable to trauma and in-

fection at birth and thereafter. Characteristically an eschar forms quickly by drying of secretions over the denuded area (Figure 103). This eschar may be very brittle and crack easily, resulting in fatal hemorrhage. It is preferable, therefore, to keep this eschar from forming by doing a primary plastic operation on the scalp, or if the defect is too large, by keeping it continuously dressed in such a way as to prevent the denuded area from drying out at any time. If such an infant is first seen after an eschar has already formed, the latter should be dissected off under general anesthesia with full preparations to manage severe hemorrhage; reformation of the eschar should be prevented until epithelization is complete (Figure 104).



Figure 105. One and a half year old child with exophthalmos on the left and protrusion of the left temporal region due to congenital absence of the lesser wing of the sphenoid bone on the left.

A congenital defect of the sphenoid bone may also occur. This was first described by Dandy [54] and subsequent cases have been reported by Le Wald [119] and Robertson.[167] Two striking examples have been seen in this clinic (Figure 105). The defect is unilateral and may involve both the lesser and greater wings and the anterior clinoid (Figure 106). Because of this, there is displacement of the orbital contents downward and forwards. In fact, the initial complaint is usually exophthalmos. There is also prominence in the fronto-temporal area. In our cases as well as Dandy's there was a large reservoir of subarachnoid fluid over the temporal pole with dilated cortical vessels in the same region. This defect has been reported by Le Wald [119] and Moore [143] in association with neurofibromatosis.

The clinical problem is a cosmetic one primarily due to the visible deformity but may be more serious if the exophthalmos is extreme or pulsating. There have been no abnormal neurological symptoms or signs in our two cases. Mental development has been normal.

If there is pulsating exophthalmos which is progressive with growth of the child, replacement of the defective orbital roof and lateral wall by a

bone graft or tantalum mesh should probably be undertaken. Lateral defects in the greater wing of the sphenoid may be repaired by tantalum cranioplasty (Figure 107).

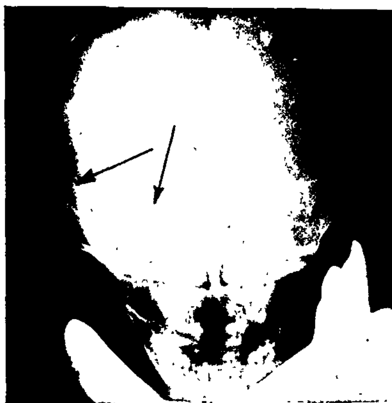


Figure 106 Antero-posterior roentgenogram of one year old girl with congenital defect of the right sphenoid bone and pulsating exophthalmos. Note the defect in the lesser wing of the sphenoid seen through the orbit and of the greater wing seen in the temporal region.



Figure 107. Congenital defect of sphenoid bone. Post-operative roentgenogram after repair of roof of the orbit with tantalum mesh and fascia and repair of temporal defect with tantalum plate. Same patient as Figure 106.

Chapter VI

Platybasia

PLATYBASIA, or basilar impression, is a developmental skeletal abnormality involving the base of the skull and the upper cervical segments of the spine. The foramen magnum is usually small, deformed and displaced upward into the cranial vault. The clivus is elevated. The atlas is underdeveloped, may be asymmetrical, is usually fused to the occiput, and the laminae may be incompletely formed. The odontoid process of the axis projects anteriorly and cranially from its normal position and invades the spinal canal. In the words of Chamberlain [29] "It is as though the weight of the head has caused the ears to approach the shoulders, while the cervical spine, refusing to be shortened, has pushed the floor of the posterior fossa upward into the brain space."

The importance of this malformation lies in the secondary effects produced on the nervous system. Although this is a congenital defect, it apparently alters as growth proceeds or at least the resultant neurological manifestations are progressive in severity. It is rare for symptoms and signs referable to this lesion to become manifest before the latter part of the second or even the third decade of life. In this clinic, one 12 year old child with rapidly progressive weakness of the legs, hyperactivity of the reflexes, sensory loss and incoordination has been treated surgically with an extremely satisfactory result (Figure 108). A number of other children have shown minimal degrees of basilar impression on x-ray examination with insufficient evidence of neurological impairment to warrant operation. We have seen adolescents come to surgical treatment on two other occasions.

The clinical appearance is characterized by shortness of the neck with a tendency toward hyperextension. The neurological manifestations are those of upper cervical cord compression: progressive weakness of a spastic variety, staggering gait, dysmetria, nystagmus, and progressive sensory loss, particularly of position and vibration senses. There may be involvement of the lower cranial nerves and there may be evidence of chronic increased intracranial pressure. The picture may simulate cervical syringomyelia, multiple sclerosis or hydrocephalus.[80, 125, 160, 179]

The diagnosis is confirmed usually by x-ray examination. This should include antero-posterior, postero-anterior, basal and true lateral views. In the basal view distortion and displacement of the foramen magnum are



Figure 108. Lateral roentgenogram of skull in 13 year old boy with platybasia. Note complete fusion of C_1 to the occiput, elevated position of the odontoid process and flattened base of the skull.

best visualized. The true lateral view demonstrates cephalic bulging of the clivus, fusion of the atlas to the occiput and cephalic projection of the odontoid process above a line drawn from the dorsal lip of the foramen magnum to the dorsal margin of the hard palate.

Treatment consists in surgical decompression of the area of the central nervous system which is involved. This amounts to essentially the same type of operative procedure outlined elsewhere (p. 45) in the treatment of the Arnold-Chiari malformation. Surgical treatment of platybasia is difficult and hazardous. It should be performed under local or preferably endotracheal inhalation anesthesia. The exposure is difficult because of the restriction in flexion of the neck. The arch of the atlas may be buried so that actually laminectomy of C_2 is necessary before C_1 can be brought into view. The angulation of the cord and dura at the cerebello-medullary junction may be markedly acute necessitating considerable care in removal of the dorsum of the foramen magnum. The dura should be opened widely and sutured back to the paraspinal muscles. A generous suboccipital decompression and removal of the arches of C_1, C_2 and C_3 is usually indicated.

These patients should be handled during the post-operative period with extreme care. They should be turned cautiously with as little motion of the neck as possible. Mechanical suction, oxygen and facilities to carry out emergency ventricular tap if necessary should always be available.

Although only one child has been operated on during the first 12 years of life here, we anticipate that more cases should be recognized and treated during this early period. It is felt that the marked disturbance of function which often comes on very abruptly in later life, as well as the high mortality when operation is performed only after extreme signs have appeared, should be reduced measurably if decompression is carried out earlier in life

PART II

HYDROCEPHALUS

Chapter I

Hydrocephalus

HYDROCEPHALUS is a pathological condition characterized by an increase in the amount of cerebrospinal fluid which is, or has been, under increased pressure, and has therefore produced an enlargement of the cerebrospinal fluid pathways. It may be found at any period of life and may be due to a variety of causes; congenital, neoplastic, post-infectious, post-traumatic, post-operative. It should be distinguished carefully from abnormal collections of fluid which are not under pressure, such as occur in porencephalic cysts within the brain substance (Figure 371) or in enlarged cerebrospinal fluid pathways that occupy the space remaining after cerebral atrophy.

Hydrocephalus is not synonymous with enlargement of the head. It must be distinguished from hereditary macrocrania, where the ventricular system is normal in size, and from chronic subdural hematoma in infancy (p. 188), where increase in head size is due to fluid in the subdural spaces. In older children and adults, the reverse may be true; that is, dilatation of the spinal fluid pathways may occur with no increase in size of the head. It should be re-emphasized, therefore, that whatever the etiology and whatever the special characteristics in each patient, the essential feature of all hydrocephalus is not head enlargement but dilatation of the cerebrospinal fluid pathways produced by an abnormal quantity of spinal fluid under increased pressure.

A wide variety of etiologic, anatomical, physiological and clinical subdivisions of hydrocephalus have been described in such a manner as to give rise to a great deal of confusion in terminology. These diverse terms will be avoided in so far as possible.

From a clinical, and particularly a therapeutic point of view, it is instructive to think of the entire nervous system as submerged in cerebrospinal fluid which is contained in a series of connected spaces. These spaces begin to make their appearance about the fifth week of embryonic life; that is, at the same time that choroidal tufts begin to be recognizable within the ependyma-lined central vesicles of the primitive brain. Weed[203] felt from his embryological studies that at this period fluid began to seep through the remaining fragment of roof plate overlying the fourth ventricle

and permeate the perimedullary mesenchymal syncytium to create these leptomeningeal spaces.

In any case, under normal conditions a continuous pathway develops which includes the ventricular system, the large cisternae at the base of the brain and the complex subarachnoid spaces covering the entire brain and spinal cord. Through this intricate reservoir cerebrospinal fluid moves, not according to the Galenic conception of ebb and flow, but principally in a definite direction. This unidirectional flow, or what Cushing termed the "third circulation",[41] results from the fact that the cerebrospinal fluid takes its origin for the most part within the lateral, third and fourth ventricles of the brain and returns to the circulating blood for the most part through the walls of the capillary bed of the pia-arachnoid covering the cerebral and cerebellar hemispheres and to a less extent the spinal cord and segmental nerve roots.

Communication between the ventricles and the surface subarachnoid pathways was first described in 1825 by Magendie[129] who discovered the mid-line foramen bearing his name and connecting the fourth ventricle with the cisterna magna cerebello-medullaris. In 1855 Luschka[128] confirmed this finding and also described openings at the apices of the lateral recesses of the fourth ventricle. In 1875, the Swedish investigators, Key and Retzius,[109] published their classic studies on the anatomy of the subarachnoid pathways, particularly the cisternae at the base of the brain. Based largely on their dissections and injection experiments, it became apparent that the large cisterna magna connected caudally with the subarachnoid space surrounding the spinal cord, and cranially by way of ventral and lateral projections with the large and intricate cisterna basalis. From the several divisions of the latter (pars pontis, pars interpeduncularis, pars chiasmatis) direct connections were traced around the brain stem with the cisterna ambiens and cisterna corporis callosi and forward with the cisterna laminae cinerea terminalis. From these cisternae, direct continuity was demonstrated with the complex subarachnoid spaces investing the cerebral and cerebellar cortex.

No attempt will be made here to summarize the data indicating that cerebrospinal fluid is formed by the choroid plexus further than paying tribute to Dandy's well-known experimental studies in the period from 1913 to 1922.[49, 50, 51, 52, 53, 57, 58] Innumerable clinical and pathological observations have confirmed the origin of the fluid from within the ventricular system and its lack of absorption at a normal rate if its egress from the ventricles is impaired Likewise, numerous clinical and pathological observations have demonstrated that obliteration of the basilar cisternae or of the surface subarachnoid pathways results in a failure of the normal rate of spinal fluid absorption.

ETIOLOGY

It is useful to think of hydrocephalus simply as the condition which results when obstruction occurs at any point in the cerebrospinal fluid pathway between the point of principal formation of the fluid within the ventricular system and the point of principal absorption in the subarachnoid spaces over the cerebral surface. Subsequent to obstruction, dilatation of the entire pathway proximal to this point occurs. The nearer to the source of the fluid, the greater the degree of dilatation usually. Common sites of obstruction seen clinically are the foramen of Monro, the third ventricle, the Aqueduct of Sylvius, the foramina of Luschka and Magendie, the cisterna magna, and the basilar cisternae.

Is there any other cause for hydrocephalus than obstruction along the cerebrospinal fluid pathway? Theoretically, over-production of fluid in the presence of a normal rate of absorption would produce the same anatomical changes. Dilatation of the ventricular system in the absence of obstruction has been seen in an occasional patient due to large adenomata of the choroid plexus of the lateral ventricle (p. 306), but aside from this there is no evidence that hydrocephalus is ever produced by hypersecretion.

Theoretically also, chronic obstruction of the venous drainage of the cerebral hemispheres, particularly by longitudinal sinus thrombosis, might result in diminution of the normal rate of spinal fluid absorption with subsequent hydrocephalus. The failure of all efforts to produce this experimentally[12] and the lack of clinical evidence of more than transient dilatation of the ventricles associated with sinus thrombosis would seem to indicate that this is a rare, if ever genuine, cause of hydrocephalus. The experimental work of Bedford[13, 14, 15] and Schlesinger[176] and the lack of any undisputed human evidence would seem to refute the possibility of hydrocephalus resulting from obstruction of the vein of Galen.

What are the causes of obstruction, then, along the cerebrospinal fluid pathway which result in hydrocephalus, and which of these are important in the pediatric age group? They may be considered under the following divisions: (1) neoplasm; (2) congenital malformation; and (3) inflammatory and post-inflammatory reaction. Reference is made to the monograph of Russell[168] for a more thorough discussion of etiology and pathology than will of necessity be presented here.

Neoplasm

Intracranial neoplasms may cause hydrocephalus by *mechanical* obstruction of any part of the cerebrospinal fluid pathway. The treatment of such hydrocephalus consists in a direct attack on the *tumor itself*, or if the tumor proves inoperable, a palliative procedure in which spinal fluid is shunted around the point of obstruction.

In infancy and childhood, hydrocephalus in this group is most commonly produced by neoplasms obstructing the fourth ventricle or the caudal opening of the Aqueduct of Sylvius (Figure 109). These tumors are usually gliomas of cerebellar or ponto-medullary origin, more rarely dermoids or choroid plexus adenomas. Gliomas of the brain stem and basal ganglia are prone to obstruct the posterior third ventricle and aqueduct as are tumors in the region of the pineal gland. In childhood, gliomas occasionally occlude the anterior portion of the third ventricle, but obstruction here is most commonly due to craniopharyngiomas. Rarely in childhood, hemisphere gliomas occlude a portion of one lateral ventricle or compress the area of the interventricular foramina so as to produce dilatation of the opposite lateral ventricle.

The clinical features and surgical treatment of neoplasms are discussed elsewhere (p. 221).

Congenital Malformations

Spina Bifida and Cranium Bifidum: The frequent association of hydrocephalus with spina bifida, and especially with myelomeningoceles, is gen-

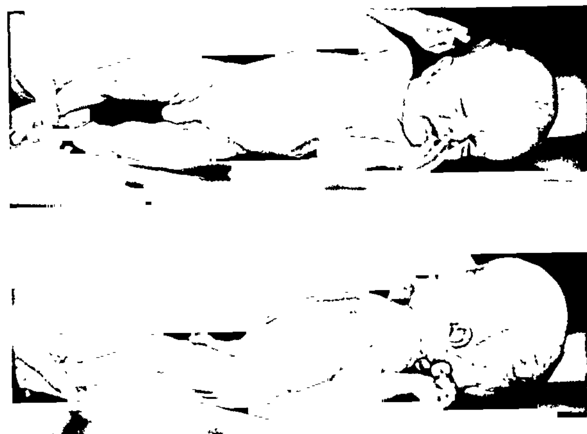


Figure 109. Two and a half month old infant with extreme hydrocephalus due to an ependymoma of the fourth ventricle.

eral knowledge. It is felt that this hydrocephalus is always related to the presence of the Arnold-Chiari malformation, wherein congenital malformation of the hind-brain results in the obliteration of the surface subarachnoid pathways around the brain stem and upper cervical cord (p. 42).

Hydrocephalus is also commonly associated with encephalomeningoceles in the suboccipital region. Obstruction to spinal fluid circulation in these cases is due to a wide variety of malformations within the posterior fossa but usually involving obliteration of the cisterna magna or the basilar cisternae at the incisura of the tentorium (p. 49).

Stenosis of the Aqueduct of Sylvius: Whether or not this common cause of hydrocephalus in early life is a congenital malformation or whether it may be inflammatory is not always clear. Russell[168] defines congenital stenosis on the basis of an aqueduct which is histologically normal but abnormally small and about which there is no increase in subependymal glia. She believes that simple stenosis of the aqueduct is rare, and that much more common is what she terms "forking" of the aqueduct in which the normal single channel may be subdivided into many minute tubules, but again with no increase in glial tissue. Patients with these lesions usually show hydrocephalus at birth or develop it rather rapidly during the first few months of life.



Figure 110. Three and a half year old boy with presumed congenital gliosis of the aqueduct. Patient presented with chronic increased intracranial pressure and signs suggesting posterior fossa tumor. Operation revealed small posterior fossa, small fourth ventricle and occluded aqueduct. No evidence of tumor.

Gliosis of the Aqueduct: This condition is distinguished from stenosis of the aqueduct by overgrowth of densely fibrillary subependymal neuroglia. This is a progressive lesion and the development of hydrocephalus is in proportion to the rapidity with which this gliosis produces aqueductal occlusion. Thus, symptomatic hydrocephalus may occur during infancy but is more apt to develop in early childhood (Figure 110) and may not produce symptoms until adolescence or early adult life. Whether this

gliosis is of congenital, neoplastic, or inflammatory origin is not clear, but from a practical point of view its progressive occlusion of the aqueduct may lead to severe and fatal degrees of hydrocephalus if not relieved.

Congenital Septa: It is possible that congenital neuroglial membranes may be present in the aqueduct or occluding the outlets of the fourth ventricle. However, it would be difficult to prove that any one of these was not the result of some inflammatory process. With congenital obstruction of the foramina of Magendie and Luschka, a tremendous cyst—which is actually the distended roof of the fourth ventricle—fills the enlarged posterior fossa, displacing the tentorium upward and compressing the cerebellar hemispheres laterally (p. 154).

Vascular Malformation: Hydrocephalus may be due rarely to congenital malformations of the cerebral vascular system. Several arteriovenous aneurysms involving the posterior cerebral arteries and the vein of Galen or the straight sinus have been reported producing occlusion of the aqueduct (p. 397).[172]

Inflammation

Hemorrhage: There is more and more clinical evidence available that intracranial bleeding at the time of birth or before birth may be followed by fibrosis of the leptomeninges, particularly at the base of the brain. This fibrosis, together with organization of residual blood within the basilar cisternae, may lead to progressive obliteration of the latter spaces.

Infection: Hydrocephalus secondary to bacterial infection of the meninges has always been a severe problem in infancy. With the advent of antibiotic therapy, the number of patients in whom a bacteriologic cure of meningitis is brought about only to be followed by obliteration of the subarachnoid pathways may be expected to increase.

Ventricular dilatation may begin during the acute phase of pyogenic meningitis if circulation becomes impaired due to actual mechanical blockage of the aqueduct or basilar cisternae by purulent exudate. More often, however, hydrocephalus is a post-meningitic process (Figure 111). Symptoms of increased intracranial pressure and gradual enlargement of the head develop at a period weeks or even months after acute infection has subsided (Figure 112). If these cases are examined pathologically, translucent thickening of the pia-arachnoid is present around the cisternae at the base of the brain or there may be virtually complete obliteration of the subarachnoid pathways around the brain stem, the optic chiasm and into the Sylvian fissures (Figure 113). Marked dilatation of the entire ventricular system including the aqueduct, fourth ventricle and possibly the cisterna magna as well is disclosed. In tuberculous meningitis, obstruction is apt to be limited to the region of the cisternae chiasmatis and interpedun-

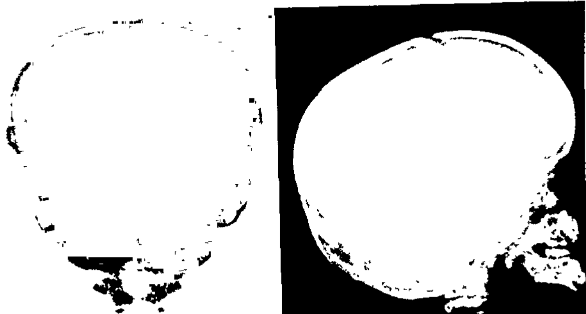


Figure 111. Antero-posterior and lateral skull roentgenograms of a three year old child with chronic, slowly progressive hydrocephalus following meningococcic meningitis shortly after birth.

cularis (Figure 114), whereas after staphylococcus, pneumococcus and meningococcus infections, it is more widespread.

It seems probable that many of the cases of so-called "idiopathic" hydrocephalus of early life, where there is no obstruction between the ventricles and the spinal subarachnoid space but an apparent failure of normal absorption, are actually post-inflammatory in origin. The incidence of meningitis in the first two years of life is particularly great and the clinical manifestations often atypical. It is probable that fetal and neonatal meningeal inflammation frequently passes undiagnosed. The increased incidence of hydrocephalus following antibiotic cure of meningitis would support the probability that this lesion also occurs after spontaneous cure. It is certainly true that careful clinical observations and complete pathological studies markedly reduce the number of cases of "idiopathic" hydrocephalus.

CLINICAL FEATURES

The clinical features of hydrocephalus are those of increased intracranial pressure. In infancy these are modified by the enlargement of the head made possible when pressure increases before the sutures have united firmly.

The classical characteristics of the hydrocephalic infant are widely recognized (Figure 115). The head appears large with respect to the body and this is confirmed by comparison of the occipito-bregmatic circumfer-

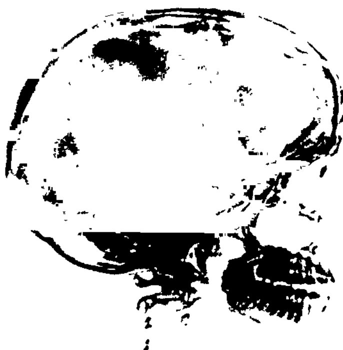


Figure 112. Lateral roentgenogram after injection of air into the ventricular system in a 13 year old girl with chronic hydrocephalus due to adhesive arachnoiditis in the posterior fossa. At operation virtually complete obstruction to the outlets of the fourth ventricle was present.

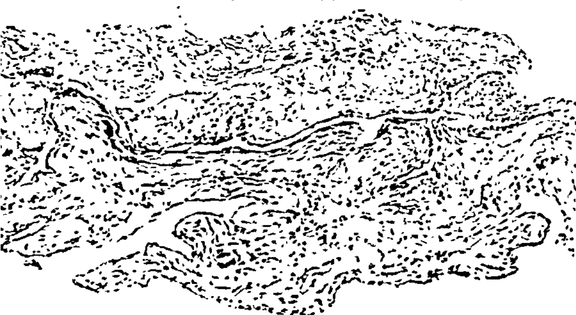


Figure 113 EMB x 200 Photomicrograph of the occlusive membrane removed surgically from posterior aspect of fourth ventricle Note the densely collagenous fibroplastic connective tissue with no evidence at this time of acute inflammation



Figure 114. EMB x 200 Photomicrograph of section through tubercle of the meninges in patient with hydrocephalus. The section presents a mass of fibrocollagen with small areas of hemorrhage, lymphocytic infiltration, Langerhans type giant cells and fibroblasts

ence with the circumference of the chest and with normal measurements for the age taken from standard charts (pp. 30, 31). More significant than any single observation are repeated measurements which indicate that the rate of growth is abnormally rapid. The disproportion of the cranial size to that of the face may be striking. The forehead is prominent, the scalp appears thin, tightly stretched and shiny, and the veins of the scalp are dilated, especially when the infant cries. The fontanelles, particularly the anterior, are enlarged, distended and tense; when the baby is placed in the upright position, the fontanelle does not become depressed and there is no pulsation visible. The sutures may be palpably separated. On percussion of the cranium, a hollow, "cracked-pot" type of note is obtained rather than the normal flat response.

The eye-balls are usually displaced downward and occasionally outward to some degree due to pressure on the thinned-out supraorbital plates. The sclera is visible above the iris (Figure 132); indeed, in severe hydrocephalus the iris may be directed downward entirely beneath the lower lid. There is also apt to be extra-ocular motor weakness resulting in various types of

strabismus (Figure 116). Random eye movements and spontaneous nystagmus are common. Examination of the eye-grounds reveals little or no papilledema usually in infants in whom enlargement of the head has occurred. In extreme degrees of hydrocephalus, however, optic atrophy may be evident.

The behavior of the hydrocephalic infant depends largely upon how adequately increase in head size is effective in compensating for increase in intracranial pressure. When the process advances slowly, there may be no disturbance of appetite, no vomiting, no retardation of motor and mental development, no hyperirritability, and few, if any, abnormal neurological findings (Figure 117). On the other hand, with increased, uncompensated intracranial pressure, there may be failure to gain, vomiting, extreme instability, unusual somnolence, disorganized motor activity, tonic neck reflexes, hyperactive deep tendon reflexes, ankle clonus, convulsions, or actual disturbance of the vital signs. Thus, an extreme degree of ventricular dilatation may be seen in one infant with very little evidence of neurological damage, and another may show marked disturbance of function, which may or may not be reversible, with much less but much more rapid ventricular enlargement. With marked dilatation of the lateral ventricles, whether rapid or slow, there is usually spasticity in the lower extremities, due presumably to stretching of the cortico-spinal tracts from the para-



Figure 115. Typical appearance of nine months old infant with severe, rapidly progressing communicating hydrocephalus.

central areas around the *dilated ventricles*. Hyperactivity in the upper extremities is apt to be less marked, due to the shorter course of the corresponding cortico-spinal tracts.[203]

DIAGNOSIS

Detection of hydrocephalus in infancy is usually not difficult because of the visual prominence of the physical signs noted above. In older children the diagnosis must be suspected in the presence of symptoms and signs of increased intracranial pressure. Confirmation of the diagnosis and more particularly of the site of obstruction to the circulation of the cerebro-spinal fluid is then made by a series of roentgenographic and laboratory procedures *(Table VIII)*.

Plain roentgenograms of the skull are first made. These confirm the increased size of the cranial vault compared to the facial bones, show separation of the cranial sutures, thinning of the bones of the vault and occasionally of the posterior clinoid processes, and soft tissue swelling in the region of the anterior fontanelle (Figure 118). Cranial defects and abnormal intracranial calcification are noted.



Figure 116. Marked internal squint secondary to chronic internal hydrocephalus due to meningococcic meningitis in infancy. Same patient as Figure 111.

Careful head measurement is made with a steel tape. The entire head is shaved and subdural taps are carried out through the coronal sutures to rule out the possibility of subdural fluid collections (p. 192). Every year several patients are seen with a diagnosis of early hydrocephalus who turn out to have bilateral chronic subdural hematomas. If the subdural taps are negative, a spinal puncture needle is introduced millimeter by millimeter through the coronal suture until the lateral ventricle is encountered. The depth at which fluid is first recovered is noted. Lumbar puncture is then carried out and simultaneous measurements of the intraventricular and spinal pressure are made (Figure 119). Samples of fluid

TABLE VIII

Date_______________________________

Name of Patient____ Age________

Head Circumference: (occipito-bregmatic)_______________________________cms.

Subdural Taps: Left_______________________ Right_______________________

Hydrodynamics: (tilt table)

	Ventricular pressure	Lumbar pressure
Horizontal	__________mms. water	__________mms. water
Head down 30°	__________mms. water	__________mms. water
Head up 30°	__________mms. water	__________mms. water

Spinal Fuid Studies:

	Ventricular fluid	Lumbar spinal fluid
White blood cells	____________	____________
Red blood cells	____________	____________
Color	____________	____________
Total protein	____________mgms %	____________mgms. %

Phenolsulphonephthalein Dye Studies:

Amount of dye injected into ventricle_______________________

Time of appearance in lumbar spinal fluid_______________________

Urinary excretion:

	Volume of urine	% of dye excreted
2 hours:	____________	____________%
12 hours:	____________	____________%

X-Ray—Ventricular Air Bubble Studies:

Depth of cortex. left____________cms. right____________cms.

Air in third ventricle____________________

Air in fourth ventricle____________________

Air in subarachnoid space____________________

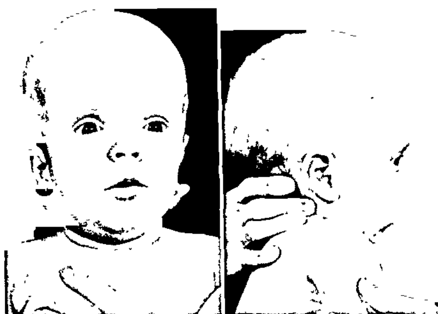


Figure 117. Three and a half months old infant with communicating hydrocephalus, slowly progressive. Contrast appearance with Figure 115.



Figure 118. Lateral roentgenogram of four month old infant with communicating hydrocephalus Note large size and thinness of cranial vault, separation of sutures and blunting of posterior clinoids.



Figure 119. Combined ventricular and lumbar puncture on tilt table with simultaneous measurement of pressure. Level of the fluid column in the manometers is compared when the table is tipped and during jugular and abdominal compression.

are withdrawn for cytologic study and protein determination. With manometers attached to both needles, a variety of manipulations may be informative. If there is free communication between the ventricles and the lumbar subarachnoid space, the pressures should be the same in the horizontal position and should vary freely and equally as the position is changed. If the infant cries or jugular compression is applied, equal rise in pressure occurs in both compartments. When there is obstruction between the ventricles and the lumbar subarachnoid space, as in aqueductal and posterior fossa lesions, the pressure in the two manometers may or may not be the same, but on jugular compression, crying, and tilting of the patient head up or down, the variations in pressure in each manometer *have little relation to one another. Also the protein and cell content of* the two fluids are apt to be quite different.

Communication between ventricle and spinal subarachnoid space is checked by introducing 1 cc. of neutral phenolsulphonephthalein into the lateral ventricle after pressure measurements are made and checking the appearance time of the dye in the fluid allowed to drip freely from the needle in the lumbar space. Normal appearance time varies from two minutes to as much as twelve. The end point is determined by pink color appearing on a gauze sponge soaked in alkaline solution. If there is no appearance of the dye in 20 minutes, an effectively complete block is assumed.

If there is any suspicion of a block between the two lateral ventricles, a needle is inserted into the ventricle of the side opposite that from injection of the dye to test for its passage from one side to the other.

injected, all urine is coll rainage device on a met dye absorbed from the subarachnoid pathway and excreted by the kidneys in two and 12 hours is measured. It is well to supplement normal fluid intake with intravenous or subcutaneous fluid to insure substantial urine output during the procedure. With a good urine output, 25 to 40% of the dye should be excreted in two hours and 50 to 70% in 12 hours in the normal subject.



Figure 120. Hydrocephalic infant on metabolism mattress for collection of total urinary output during phenolsulphonephthalein excretion studies. Glass funnel over genitalia connected by rubber tube through hole in mattress to drainage bottle.

(1) If there is appearance of dye in the lumbar fluid in two to six minutes but only 8 to 15% of the dye is excreted in 12 hours in the urine, it means extreme obstruction of the surface subarachnoid pathway at a point distal in the circulation of spinal fluid to the cisterna magna.

(2) If the dye appears promptly in the lumbar fluid and 20 to 30% is excreted in the urine in 12 hours, it means partial obliteration of the surface subarachnoid pathways.

(3) If no dye appears in the lumbar fluid, a block within the ventricular system, aqueduct or cisterna magna is assumed. Under these circumstances, there is usually a very small percentage of dye excreted in the urine—less than 10% in 12 hours.

Ventriculographic studies usually complete the investigation. Infants with marked hydrocephalus from whatever cause do not tolerate encephalography well and, of course, in the presence of posterior fossa obstruction it may be extremely dangerous. When there is marked intracranial pressure with extreme dilatation of the ventricles in young babies, complete replacement of fluid by injection of air directly into the ventricles is also

poorly tolerated. In this group of patients, however, a ventricular "bubble" study is safe and usually gives the information desired. Thirty to 50 ccs. of cerebrospinal fluid are removed from one lateral ventricle and replaced by air or ox are made in several positions; antero-posterior postero-anterior, lateral, brow-up lateral, occiput-up lateral, and upside down. The thickness of the cortex can be estimated adjacent to the part of the ventricle uppermost in each view (Figure 121). In the upside down and occiput-up films, it is usually possible to demonstrate air in the fourth ventricle, cisterna magna and even the spinal subarachnoid obstruction to the outlets of th oiditis these views show an enlarged fourth ventricle but no air distal to this point (Figure 123). It should be emphasized here that if this study is unsatisfactory, or there is any question of a supratentorial mass lesion, more complete ventriculography should be done in the usual manner.

Accurate visualization of the size of the posterior fossa may be obtained by dural sinograms. This is helpful in differentiating stenosis of the aque-

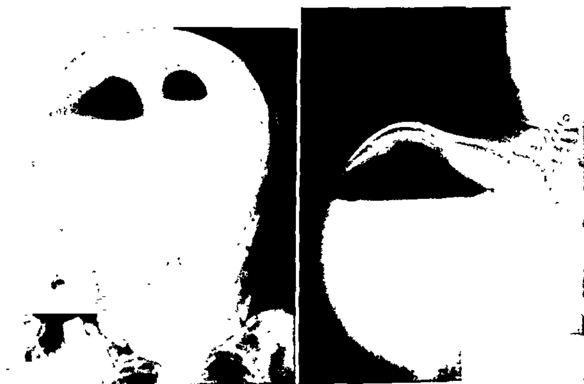


Figure 121 *Ventricular "bubble" study in four weeks old baby with congenital obstruction of the Aqueduct of Sylvius; 40 ccs. of air injected into the right lateral ventricle.*

A. Antero-posterior upright film.

B. Upside down film. No air has passed into the aqueduct or fourth ventricle. *Note small size of the posterior fossa.*



Figure 122A. Upside down lateral ventricular air study showing air in the cisterna magna and in the spinal canal.

B. Upside down lateral ventricular air study showing air in the aqueduct, dilated fourth ventricle, and cisterna magna.

In each instance approximately 40 ccs. of air were introduced into a lateral ventricle through a needle passed through the coronal suture.

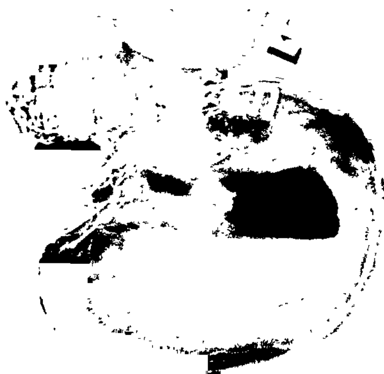


Figure 123. Upside down lateral ventriculogram of five year old child with marked hydrocephalus due to adhesive arachnoiditis obstructing the outlets of the fourth ventricle (see Figure 113). Note marked dilatation of the fourth ventricle as well as *the third and* lateral ventricles, but no air escaping into the cisterna magna or surface pathways

duct (Figure 124) from obstruction of the outlets of the fourth ventricle (Figure 139).

TREATMENT

There are few pathological conditions that have received more intensive, more diverse, more desperate attempts at treatment than the hydrocephalus of infancy.[59] The obvious physical deformity and progressive neurological deterioration of these pitiable children has challenged the medical profession from its beginnings to seek a permanent cure. Real progress toward this goal has been furthered principally by knowledge of the anatomy of the subarachnoid pathways, an ever-increasing understanding of the formation, circulation, and absorption of cerebrospinal fluid and by the remarkable technical advances in neurological surgery of the past 50 years.

Historical attempts at treatment have included: (1) simple puncture of the lateral ventricle; (2) blood-letting; (3) diuresis; (4) use of gland extracts; (5) dehydration; (6) tight bandaging of the head; (7) roentgen treatment of the choroid plexus; (8) resection of the choroid plexus; (9) ligation of the carotid arteries; (10) lumbar puncture; (11) constant ven-



Figure 124 Antero-posterior and lateral roentgenograms after injection of diodrast into the longitudinal sinus at the anterior fontanelle. Appearance is characteristic of hydrocephalus due to stenosis of the Aqueduct of Sylvius. Note extremely small size of the posterior fossa as outlined by dye in transverse sinuses Compare with Figure 139

tricular drainage to the surface of the body, the subaponeurotic tissues, the subarachnoid and subdural spaces, the cerebral veins, the longitudinal sinus, the scalp and neck veins, the orbit, nasopharynx, paranasal air sinuses and mastoid; and (12) constant spinal drainage to the subcutaneous tissues, the peritoneum and retroperitoneal space, the bowel, bone-marrow and the urinary tract. The innumerable published reports of these varied treatments are for the most part a recital of discouragement and failure. Frequent individual patients appeared to respond well temporarily to some new surgical procedure only to have failure ensue later. However, the problem is by no means hopeless, and advances in this field since World War II have been extremely encouraging.

It should be remembered that hydrocephalus is not a single disease; it is a pathological condition which may be due to numerous different

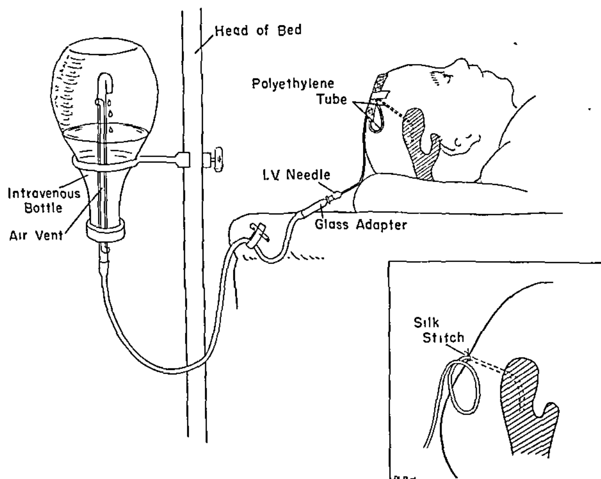


Figure 125. Constant Ventricular Drainage.

A plastic catheter is inserted into the ventricle through the coronal suture or a small drill hole. Drainage bottle is attached to the *head of the bed*. Height of the inverted "J" tube within the bottle above the level of the lateral ventricle determines the level at which ventricular pressure is maintained. The amount of fluid put out in any given period at any particular pressure can easily be measured.

structural abnormalities secondary to a variable etiology. The first step in treatment, therefore, is investigation of the type of cerebrospinal fluid obstruction, as outlined in the previous section.

The treatment of hydrocephalus is surgical. No medical or radiological treatment is of avail except in so far as obstruction may be due to a proven radio-sensitive neoplasm. When cerebrospinal fluid obstruction is due to a mass lesion that is accessible to direct attack, this is the obvious method of treatment. When obstruction is due to occlusion of the outlets of the fourth ventricle or of the cisterna magna by localized congenital or post-inflammatory lesions, the obvious treatment again is direct attack. Numerically, however, the great majority of patients with hydrocephalus in early life do not fit into these groups.

The methods of surgical treatment which have proved of value or are in current use in this clinic will be presented in some detail. They include: ventricular drainage, ventriculo-cisternostomy (Torkildsen), third ventriculostomy, choroid plexectomy, arachnoid-ureterostomy, ventriculo-ureterostomy, and ventriculo-peritoneostomy.

Ventricular Drainage

The indications for ventricular drainage in hydrocephalus are obviously limited to those situations in which temporary reduction in pressure is needed. This has proved of value during the pre- and post-operative periods of myelomeningocele excision in the presence of mild hydrocephalus. Also, patients acutely obstructed by an inflammatory lesion such as cerebellar abscess, or by pyogenic or tuberculous meningitis may benefit from intermittent or regulated constant ventricular drainage (Figure 125) until normal circulation of cerebrospinal fluid is re-established.[16] The same situation may be true when temporary obstruction is due to edema around the aqueduct or fourth ventricle after excision of tumors.

Ventriculo-Cisternostomy (Torkildsen)

It is the feeling in this clinic that when preliminary studies of a hydrocephalic infant or child disclose obstruction within the third ventricle or Aqueduct of Sylvius which is inaccessible to direct attack, the procedure of choice is posterior fossa exploration with preparation to perform a ventriculo-cisternostomy of the type devised by Torkildesen.[195, 196] In the pediatric age group this operation is most often performed for stenosis of the aqueduct (Figure 126) and for tumors involving the aqueduct and third ventricle, such as craniopharyngioma, pineal tumors, and gliomas of the basal ganglia and brain stem. If the cisterna magna is patent, this procedure has proved preferable to subfrontal or subtemporal third ventriculostomy.

In older infants and in children, the operation has carried no particular



Figure 126. Lateral air injection of patients with aqueductal stenosis showing rubber Torkildsen tube in position. This patient had a Torkildsen Procedure first performed in 1939. This was revised a year later and has been functioning satisfactorily now for over 12 years.

problems and is performed according to the technique described by Torkildsen. In young infants, however, particularly during the first three to six months of life, the operation is more difficult and the results are not as satisfactory. The reason for this may be that because the flow of cerebrospinal fluid is impaired by aqueductal obstruction at such an early age, the basilar cisternae and surface subarachnoid pathways simply have never contained an adequate quantity of fluid and therefore have failed to develop in a normal fashion. Consequently, even if fluid is successfully shunted from the lateral ventricle to the cisterna magna, its free circulation beyond this point is impeded by failure of the surface pathways to open up. In early infancy the operation is also somewhat more difficult technically because the dura covering the posterior fossa is poorly developed and apt to contain a continuous lake of venous blood rather than the discrete venous sinuses normally present.

Either a polyethylene or rubber catheter may be used for the shunt. In this clinic, rubber has recently been used exclusively. If interventricular obstruction is present, as with an anterior third ventricular lesion, bilateral

Torkildsen procedure is essential. This has been performed in patients with large suprasellar cysts satisfactorily (Figure 272).

The Torkildsen procedure in childhood is carried out as follows. The patient is placed in the face-down position under endotracheal general anesthesia. A small burr hole is made low in the occipital region about 2 cms. from the mid-line usually on the right side, and the dura opened

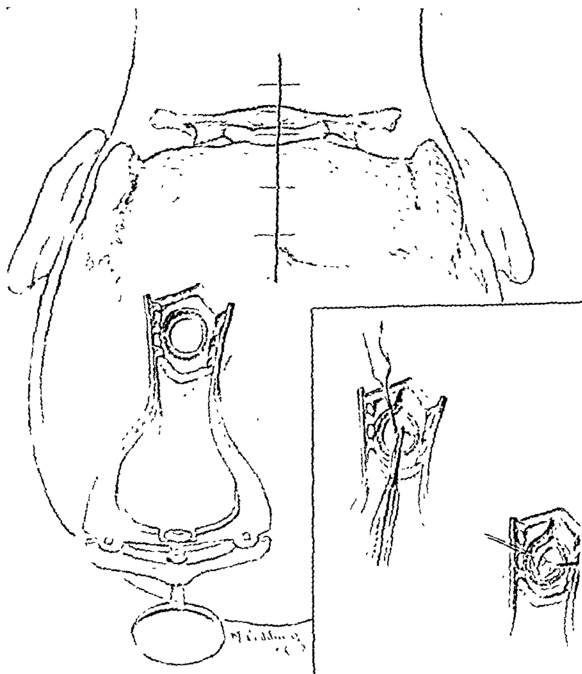


Figure 127. Torkildsen Procedure No. 1.

Position of burr hole and posterior fossa incision. Insert shows channel in bone made to receive the tube and linear incision in the dura with sutures in the dural margin in order to secure the tube at this level.

(Figure 127). A ventricular needle is introduced into the occipital horn of the lateral ventricle and along the same tract approximately 5 to 7 cms. of the catheter is introduced into the ventricular lumen. Sutures previously placed on either dural margin are then tied around the tube snugly (Figure 128).

Mid-line exposure of the posterior fossa is then carried out in the ordinary manner. A small suboccipital decompression is performed; it is usually unnecessary to remove the arch of the atlas.

Linear incision of the dura over the cerebellar hemisphere is carried diagonally toward the foramen magnum until the cisterna magna comes into view. In young infants it may be necessary to place a row of silver clips along both margins of the dural opening to control bleeding from venous lakes in the dura. If posterior fossa exploration is not contemplated, it is necessary only to visualize enough of the cisterna magna so that it can be opened. Indeed, it is preferable not to open the cistern widely but to pass the tube through a small puncture hole in its wall. If the posterior fossa is to be explored, the dura is opened widely, as well as the arachnoid of the cisterna magna, to permit exposure of the fourth ventricle.

The catheter is ordinarily passed from the burr hole wound to the posterior fossa in a plane between the galea and the pericranium (Figure 128). It may also be passed between the bone and dura. The distal end of the tube should lie in the mid-line well within the cisterna magna, not under tension, and not kinked at any point. It may also be placed in the lateral cistern, particularly if mid-line structures have been disrupted by exploration of the fourth ventricle.

When the tube is satisfactorily placed beneath the arachnoid, it is anchored in this position and the dura approximated as accurately as possible. Before closing the operative wounds, a No. 27 hypodermic needle may be introduced into the tube at a convenient point to check its patency in both directions by injection of saline (Figure 129).

Post-operatively, daily lumbar drainage of spinal fluid is performed for several days and ventricular puncture is avoided unless increased pressure cannot be otherwise relieved. Occasionally the shunt does not seem to work for several days and then suddenly free communication between the ventricle and the lumbar spine becomes established.

Third Ventriculostomy

Prior to introduction of the Torkildsen procedure in 1939, the operation in most common use for treatment of obstruction of the aqueduct or posterior third ventricle was third ventriculostomy. This operation was first performed with any success by Dandy[53] who used the subfrontal approach in the beginning and then changed to the subtemporal. Although this



Figure 128. Torkildsen Procedure No. 2

Plastic or rubber tube is passed subgaleally from burr hole incision to posterior fossa incision. The tube is introduced into the lateral ventricle along the tract made by insertion of a ventricular cannula

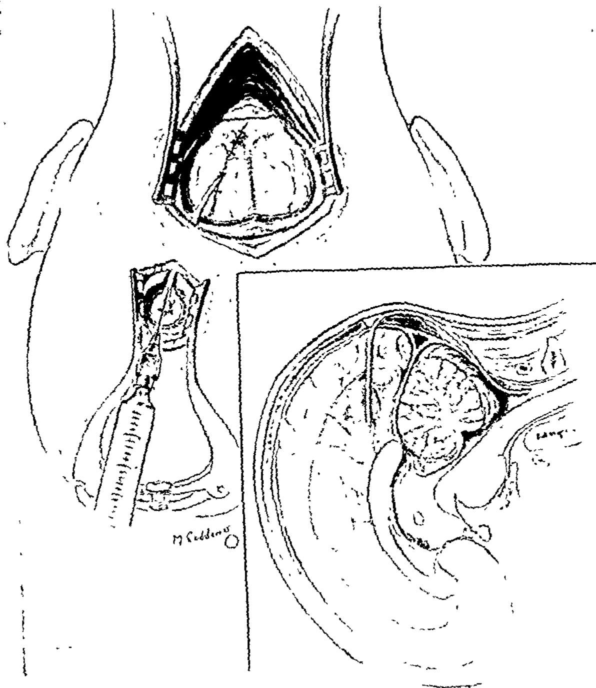


Figure 129. Torkildsen Procedure No. 3.

Irrigation of tube through hypodermic needle after the tube has been secured at both ends. Insert shows course of the tube from the lateral ventricle to the cisterna magna in longitudinal section.

operation has been abandoned for the most part, there is an occasional patient in whom the cisternae of the posterior fossa are not available for implantation of a Torkildsen tube, but the basilar cisternae of the middle fossa are still patent.

Third ventriculostomy in the first few months of life is apt to be unsuccessful, probably because the cisternae and surface pathways have never contained enough fluid to develop properly. Under these circumstances, the opening in the third ventricle usually seals over within a short time. The other common difficulty with this operation is accumulation of fluid in the subdural rather than the subarachnoid space.

Subfrontal third ventriculostomy is carried out by opening the anterior projection of the floor of the third ventricle into the cisterna chiasmatis. This region is approached by elevating the right frontal lobe after performing a small frontal craniotomy. Because it is necessary to open the shallow chiasmal cistern first and then the bulging third ventricle at a point where it is in close apposition to the floor of the skull, fluid quickly pours into the subdural space. This can be avoided to some extent presumably by the maneuver advocated by Stookey and Scarff [174, 190] of also opening the floor of the third ventricle farther posteriorly into the cisterna interpeduncularis in the region of the tuber cinereum.

Subtemporal third ventriculostomy is carried out by opening the floor of the third ventricle laterally into the interpeduncular cistern. After making a small subtemporal decompression with the patient's head in the lateral position and slightly dependent, the temporal lobe is retracted until the cisterna interpeduncularis is visualized and opened adjacent to the occulomotor nerve. The floor of the third ventricle is then opened as widely as possible where it bulges into the cistern. It is necessary to decompress the ventricular system first in order to get room to perform this procedure. Obviously, third ventriculostomy is not appropriate when there is marked obstruction of the third ventricle itself.

Choroid Plexectomy

This operation is designed to decrease the amount of spinal fluid formed within the ventricular system. Until recently it has been the only operation which has met with any success in hydrocephalus due to obstruction of the basilar cistern or surface subarachnoid pathways. Dandy [49] originally carried out this procedure by excision and later by coagulation of the choroid plexus in an air medium after removing all of the spinal fluid from the ventricle through a hollow instrument resembling an operating cystoscope.[55] This was modified by Putnam,[163] Scarff [175] and others so that coagulation of the glomus of the plexus was carried out without removing the ventricular fluid by introduction of a lighted solid ventriculoscope carrying two closely

approximated electrodes. This instrument is introduced through small occipital openings; a bilateral operation is usually carried out at one sitting.

For many reasons, including poor visibility, inadequate removal of the plexus, post-operative hemorrhage and poor exposure in case of operative difficulties, this method has been abandoned in recent years in favor of open operation, that is, transcortical exposure of the atrial region of the lateral ventricle. One side only is done at a time. These operations may be performed under local but preferably under endotracheal ether anesthesia. The baby is placed on the operating table in the lateral position with the head rotated slightly forward of the lateral position. A 6 cm. linear incision is made in the posterior temporo-parietal region (Figure 130). A one inch trephine button is removed or a single burr hole made and a circular piece of bone, one to one and a half inches in diameter cut out with stout scissors and the dura opened.

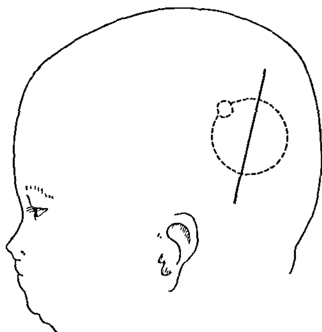


Figure 130. Position of exposure for removal of the choroid plexus from the lateral ventricle. Heavy solid line indicates the skin incision. Dotted lines indicate the extent of bone removed. This can be accomplished with a large trephine or cutting a circular flap from an initial burr hole.

After superficial cauterization, a 2 cm. cortical incision is made and dissection carried down through the thinned out white matter along the track of a ventricular puncture until the ventricle is encountered. It may be possible to expose the choroid plexus of both lateral ventricles if there is a large perforation of the septum pellucidum. The glomus of the choroid plexus is grasped gently with forceps, and silver clips are placed across the major vessels at the base. The intervening mass of the plexus is then excised. Any bleeding encountered is asily managed with the cautery or additional clips under direct vision. The cautery is used as little as possible, particularly within the body of the lateral ventricle. When as much plexus as possible has been removed and hemostasis achieved, the packs are removed and the ventricle filled with warm normal saline solution. Post-operative ventricular punctures are carried out only as indicated by bulging of the fontanelle and the baby's clinical course. If there is indication for a bilateral operation, the second side can usually be operated upon within a week to 10 days in the same fashion.

Choroid plexectomy must necessarily be a partial operation. It is impossible to remove the plexus from the third ventricle and improbable that it could very often be removed safely from the fourth ventricle in hydrocephalic infants. It seems probable that most of the favorable results from this procedure have been in patients in whom spontaneous arrest of hydrocephalus was imminent or in whom the circulation and absorption of cerebrospinal fluid were impaired to a minor degree only. It is indicated in patients with the Arnold-Chiari malformation when the head continues to grow slightly after suboccipital and upper cervical decompression. It is perhaps indicated in border-line cases of communicating hydrocephalus in whom the process seems to be abating but not completely arresting spontaneously. If there is poor absorption and excretion of a tracer dye (less than 15 to 25 per cent) from the subarachnoid space, choroid plexectomies may slow down the rate but will not halt the progression of hydrocephalus. The operation is now rarely performed in this clinic.

Arachnoid-Ureterostomy

The alternative approach to choroid plexectomy when obstruction to cerebrospinal fluid circulation exists in the basilar cisternae or surface pathways is to divert fluid from proximal to the point of obstruction either directly into the circulating blood, onto a surface sufficiently extensive and sufficiently vascular to permit absorption at a rate comparable to that in the normal subarachnoid spaces, or to divert it onto an excretory surface and eliminate it from the body. Based upon considerable laboratory experience in the diversion of spinal fluid through various types of channels in experimentally hydrocephalic animals,[93] a modification of the method of Heile[84] was devised and first put into clinical use in 1948.[132] This consists in shunting cerebrospinal fluid through a small calibre polyethylene tube from the spinal subarachnoid space into the ureter after removal of one kidney. This [illegible] and to date has given the [illegible] with marked hydrocephalu[illegible]a. Recently it has been performed on all patients with this type of hydrocephalus who seemed salvageable and in whom the hydrocephalus is unquestionably still progressing. Preliminary studies in addition to those already outlined which establish the type of hydrocephalus include intravenous urography to be certain two normal kidneys are present, urine analysis to rule out urinary tract infection, and non-protein nitrogen determinations to insure normal renal function.

The operation is performed as follows (Figure 131). Under endotracheal ether anesthesia the patient is placed on the operating table in

the lateral position with the kidney bench elevated. A subcostal incision is made, the muscles of the flank divided in turn, the false renal capsule opened after reflecting the peritoneum medially and the kidney mobilized. The ureter is identified and divided at its junction with the renal pelvis. The vessels of the renal pedicle are divided and the kidney removed. The left kidney has usually been selected since it is somewhat higher and on the side of the aorta rather than the vena cava.

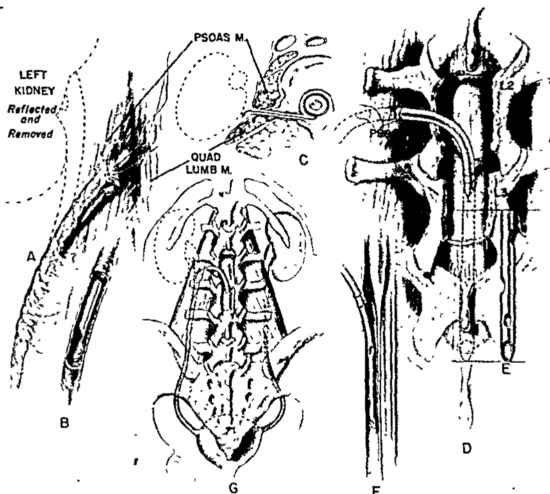


Figure 131. Arachnoid-Ureterostomy.

A. Detail of suture of the upper end of ureter to the fascia of the psoas muscle.

B. Detail of the plastic tube lying within the lumen of the ureter.

C. Cross section showing pathway of the tube from the spinal canal to the retroperitoneal space.

D. Laminectomy at L_2 and L_3, showing position of tube after insertion.

E. Detail of the end of the plastic tube inserted into the subarachnoid reservoir.

F. Relation of the tube to the dura and cords of the cauda equina.

G. General diagram of the completed operation.

Leaving the nephrectomy wound open, a short mid-line incision is made over the upper lumbar spine with the patient still in the lateral position, but with the kidney bench now lowered. A laminectomy of L_2 and L_3 is performed. A mid-line incision is made in the dura approximately 5 to 7 mms. in length in such a fashion as not to open the arachnoid. Silk sutures are tied into either dural edge. A pin point hole is then made in the arachnoid and a polyethylene tube inserted through it as spinal fluid pours forth. The tube is introduced caudally for a distance of about 4 to 5 cms. so that it floats freely among the cords of the cauda equina. Before inserting the tube, it is bent into a gentle curve at the appropriate point by dipping it in very hot saline and allowing it to cool in the proper position.



Figure 132. Four and a half months old infant one week after arachnoid-ureterostomy. Note depression of anterior fontanelle, overlapping of coronal suture and persistent downward displacement of the eyes. This child's head became approximately 3 cms. smaller in circumference following operation.

The tube is anchored by the sutures previously placed in the dural margins and the dura closed snugly around it by additional silk sutures. The tube is tunnelled through the paraspinal muscles in a smooth curve to enter the perinephric space and be inserted for a distance of 4 to 6 cms. into the lumen of the ureter. Care is taken to mobilize the ureter in such a way as to leave its nerve and blood supply intact. Three or four sutures in the margin are used to pull the ureter over the polyethylene tube and secure it firmly to the fascia of the paraspinal musculature. Ligatures are never placed around the ureter. At various times during the operation, patency of the system can be tested by having the anesthetist exert pressure on the anterior fontanelle.

Post-operatively the patients do well but need parenteral fluids for two or three days. There may be a transient rise in N.P.N. to 50 to 60 mgms. per cent. The circumference of the head in infants decreases in size from 1 to 4 cms. as the fontanelles become depressed and the sutures overlap (Figure 132). Meningitis secondary to urinary infection has been a rare complication. The plastic tube is well tolerated. Within several days of

operation, these infants begin to eat well, smile and play; they lose their hyperirritability, disorganized motor activity and restlessness.

Infants who are still largely on a milk diet, with its low sodium content, should have two to three grams of ordinary table salt (NaCl) added to the diet daily. Because of the loss of spinal fluid in the shunt, there is a continual loss of sodium from the body. In small infants this may be sufficient to bring them to the edge of negative sodium balance and any additional loss of sodium, as by vomiting, excessive perspiration, diarrhea, or lack of normal intake with intercurrent illness may precipitate acute dehydration if additional salt is not given. Older children and adults eating normally salted table food probably have an ample margin to compensate for loss through the shunt.

URETERO-ARACHNOID SHUNTS:		
PATIENTS OPERATED UPON		47
" LIVING	36	
" DEAD	11	
" REOPERATED		5
CAUSE OF DEATH:		
ACUTE DEHYDRATION	6	
INFECTION	2	
TUMOR	1	
ASPIRATION	1	
UNKNOWN	1	

Figure 133. *Analysis of results of arachnoid-ureterostomy in patients treated from 1948 to* date.

Following this procedure, the intracranial pressure remains normal or low at all times, and normal brain development is allowed to proceed. In this clinic, 47 * of these procedures have been performed (Figure 133). Two patients died early in our experience, six to 10 weeks after operation, before we realized the need for added salt to prevent acute dehydration associated with intercurrent illness. Four additional children have died from six weeks to eight months after operation due to parental neglect of dehydration occurring with gastro-intestinal infections. Two patients died in whom the operation should not have been undertaken because of the presence of an intracranial inflammatory process still active. One patient, otherwise doing well, died of aspiration pneumonitis; one died of tumor and one of causes unknown. The other 36 patients are all home and doing well. The most encouraging feature is that they are not simply kept alive by this procedure, but their development has appeared to proceed normally as nearly as clinical and psychometric evaluation is reliable. A number of infants have now been followed more than three years (Figures 134 and 135). In five patients the catheter has become obstructed in its intrathecal portion after a number of months. When the tube was replaced, they again became asymptomatic.

It is felt that most patients with this type of hydrocephalus develop secondary pathways for circulation eventually and that the need for the

* Series now includes 58 patients, of whom 41 are living and asymptomatic.



Figure 134. Arachnoid-Ureterostomy.

A (*Left*). Four and a half months old infant with all of the clinical features of extreme internal hydrocephalus.

B (*Center*) Ventricular "bubble" study of same patient showing degree of ventricular enlargement prior to operation.

C (*Right*). Same patient three years and five months following arachnoid-ureterostomy The head is still smaller than it was pre-operatively. Mental and physical development only slightly behind schedule, if at all



Figure 135. Arachnoid-Ureterostomy

A (*Left*) Appearance at four months of age Large head, irritable, strabismus, one generalized convulsion.

B (*Right*) Brow-up lateral ventricular "bubble" study showing extent of communicating hydrocephalus

tube probably lasts only a few months to a few years. If this is true, possible obstruction of the tube due to the rapid growth of later childhood and adolescence will not be important.

Ventriculo-Ureterostomy

If there is obstruction not only of the basilar and surface subarachnoid pathways, but of the posterior fossa and spinal subarachnoid spaces as well, then arachnoid-ureterostomy of the type just described is obviously impossible. This situation is encountered usually secondary to generalized meningitis. Choroid plexectomy may be helpful if circulation is only partially impaired, but if obstruction is extensive or if hydrocephalus is progressive after plexectomy, then a shunt of some kind directly from the ventricular system is indicated. Experience in the laboratory with attempts to divert fluid into the pleural cavity, thoracic duct, longitudinal sinus, and veins of the neck discouraged clinical trial of these locations. Nosik[149] has described ventriculo-mastoidostomy with successful reduction of intracranial pressure. This procedure has not been employed in our clinic.



Figure 135C. Appearance three years and two months after arachnoid-ureterostomy. This child actually seems slightly advanced.

Ventriculo-ureterostomy has been performed 11 times in this clinic.[134] In this procedure, a long polyethylene tube is inserted into a lateral ventricle for 5 to 7 cms. through a burr hole in the low occipito-temporal area (Figure 136). The tube is anchored at the dural margins and to the galea. It is led subcutaneously through the posterior neck and down the paraspinal region by passing it from one small skin incision to another. A nephrectomy is performed in the same manner as described for arachnoidureterostomy. The tube is tunnelled through the paraspinal muscles from a point just below the twelfth rib to the perinephric space and inserted into the lumen of the ureter for 4 to 6 cms. The ureter is drawn over the tube and sutured

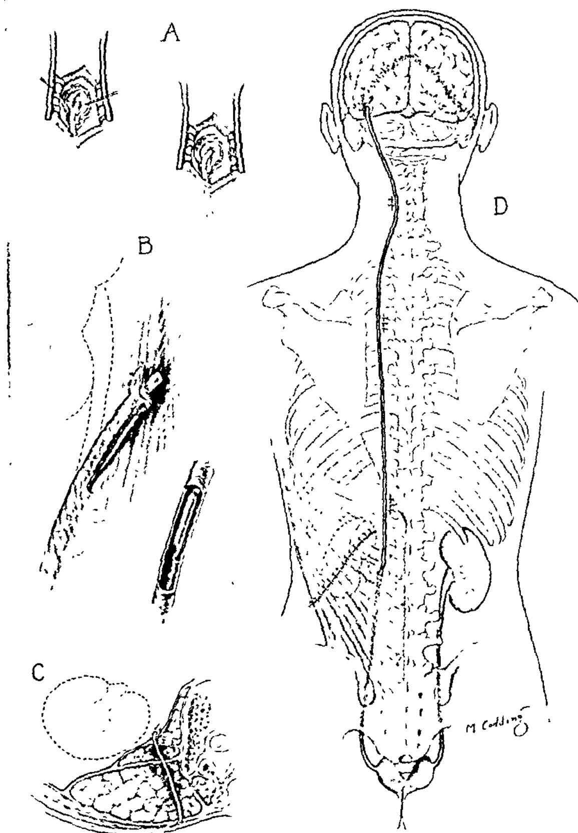


Figure 136. (*See legend on facing page.*)

to the fascia of the paraspinal musculature. With this procedure in children, one must obviously face the problem of growth. If the subarachnoid pathways do not open up as growth proceeds, the tube will certainly need to be replaced periodically. There has been one postoperative death. Three patients have had to have the tube replaced after 12 to 20 months because of it becoming too short. In another patient, small amounts of bleeding have occurred into the ventricular system and bladder unaccompanied by other symptoms. The increased frequency of bacteriologic cures of generalized pyogenic meningitis with the aid of antibiotics may result in a greater number of patients with secondary hydrocephalus necessitating surgical relief of this type (Figure 137).

In three patients with obstruction of the Aqueduct of Sylvius in early life a Torkildsen procedure has been performed to by-pass this obstruction, only to find a block in the basilar cisternae as well. In these patients, hydrocephalus progressed but was now of the communicating variety. Arachnoid-ureterostomy was therefore performed, in each instance with a successful result to date.

Ventriculo-Peritoneostomy

Diversion of cerebrospinal fluid directly from the lateral ventricle into the peritoneal cavity through a plastic or rubber tube, as first performed by Cone, has been carried out successfully by a number of neurosurgeons. Our experience has apparently been similar to others that in some patients it works well immediately but subsequently the peritoneal end of the tube becomes obstructed and is difficult to make effective again. However, in many patients the ventriculo-peritoneal tube has continued to work satisfactorily up to periods of several months. In our hands this procedure has not been as effective as the ventriculo-ureteral shunt. We now use a rubber tube in the ventricle through an occipital burr hole and anastamose it to a polyethylene tube subcutaneously mid-way down the back. The plastic tube is then introduced into the pelvic peritoneal cavity through a small, right lower quadrant muscle-splitting incision. This operation has been done principally in patients with hydrocephalus due to the Arnold-Chiari malformation in whom drainage via the urinary tract was contra-indicated. This operation has the same disadvantage as ventriculo-ureterostomy with

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Figure 136. Ventriculo-Ureterostomy.

A. Detail of burr hole and secure attachment of tube to the dura.

B. Detail of insertion of tube into the ureter.

C. Pathway of the tube from subcutaneous to retroperitoneal space.

D. General plan of ventriculo-ureterostomy.

respect to growth. The tube should probably be lengthened after 12 to 15 months unless there is evidence of spontaneous arrest of the hydrocephalus.

Lumbar arachnoid-peritoneostomy has also been carried out by a number of neurosurgeons. Our early experience with this was discouraging and arachnoid-ureterostomy was substituted because of its greater success technically. The only lumbar arachnoid-peritoneostomy performed recently, however, is working quite satisfactorily after three months, as demonstrated by rapid passage of pantopaque from the lumbar subarachnoid space through the tube to the peritoneal cavity.

The adoption of these radical surgical procedures in recent years in the treatment of hydrocephalus has so far been extremely rewarding. An attempt is made to go ahead with an operation which will reduce intracranial pressure to normal and maintain it there as soon as it is established





Figure 137A (*Left*). Brow-up lateral ventricular air study showing severe non-communicating hydrocephalus secondary to *b coli* meningitis in three month old infant.

B (*Right*). Same patient one year and nine months following ventriculo-ureterostomy. Asymptomatic, extremely active and developing

that hydrocephalus is progressive. Temporizing operations should be avoided. It is a great tragedy to keep a child alive by palliative procedures which, though preserving vital functions, do not allow cortical development to proceed at a rate adequate to prevent severe mental and physical retardation. There is, in addition, a tremendous psychological and social advantage to procedures such as these which halt at once abnormal expansion of the head.

Obviously, final judgement on this type of surgery must await the results of much longer follow-up observations. The results to date appear to warrant continuation of shunting procedures until something better becomes available.

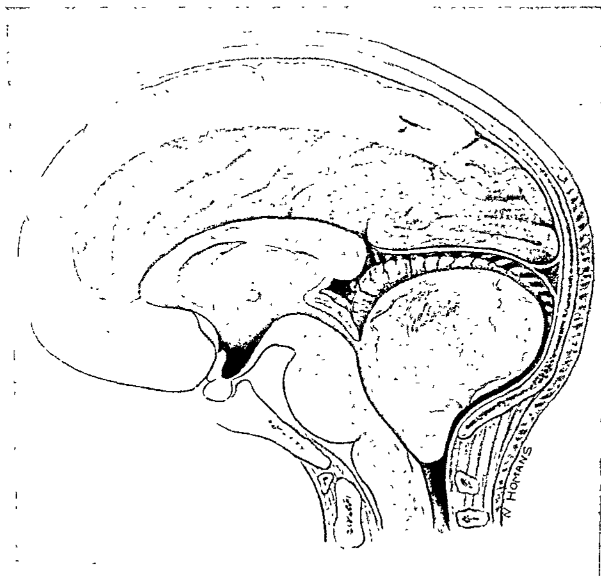


Figure 138. Congenital obstruction of the foramina of Magendie and Luschka. Sagittal section showing dilatation of fourth ventricle to form huge cyst within the posterior fossa. Note: (1) upward displacement of tentorium and torcula, (2) compression of the cerebellum, and (3) marked internal hydrocephalus.

ATRESIA OF THE FORAMINA OF MAGENDIE AND LUSCHKA

One of the more interesting and unusual types of hydrocephalus in early life deserves special comment. This is hydrocephalus due to congenital atresia of the foramina of Magendie and Luschka. If the proper diagnosis is made early enough, before irreversible neurological changes have occurred, surgical treatment is quite simple and effective.

Because of partial obstruction to the outlets of the fourth ventricle, there is symmetrical dilatation not only of the lateral and third ventricles and aqueduct, but also tremendous enlargement of the fourth ventricle itself. This results in formation of a huge cyst-like structure in the posterior fossa which displaces the cerebellar hemispheres to either side, causes agenesis of the cerebellar vermis, marked elevation of the tentorium and obliteration or failure of development of the cisterna magna (Figure 138).

As pointed out by Taggart and Walker[193] the transverse sinuses are located much higher than usual in this condition. This has been confirmed in our clinic by longitudinal sinus phlebography (Figure 139) and is a manifestation of enlargement of the posterior fossa during embryonic development. Pre-operative plain x-ray films of the skull may show the ele-



Figure 139 Antero-[illegible] months old infant with congenita[illegible] wing injection of diodrast into [illegible] e extremely high position of the [illegible] d after the posterior fossa cyst had been removed. There is still residual air in the markedly dilated ventricular system Compare with Figure 121

vated position of the torcula and transverse sinuses.[51] Combined ventricular and lumbar taps and ventricular air studies show a non-communicating type of hydrocephalus. Air in the huge posterior fossa cyst may be identified (Figure 140).

Treatment consists in exposure of the posterior fossa with removal of the cyst which presents in the mid-line when the dura is opened. Actually when the cyst is entered, it is apparent that its floor is the floor of the fourth ventricle and that one can look directly through a much dilated aqueduct into the third ventricle. The cerebellar hemispheres are displaced laterally. It is a simple matter to resect all of the cyst that is not closely attached to the cerebellum. This leaves a wide open communication from the fourth ventricle into the subarachnoid surface pathways within the posterior fossa and spinal canal. If the basilar cisternae are patent, spinal fluid circulation can then proceed normally and the hydrocephalus is completely relieved.

The end results in the cases collected and reported by Taggart and Walker were poor. However, the average age in their patients was one year, so that they must have had chronic increased intracranial pressure for a long time before any treatment was started. If the diagnosis can be



Figure 140. Upside down lateral roentgenogram after injection of 30 ccs. of air into the right lateral ventricle in a four months old infant with congenital obstruction of the foramina of Magendie and Luschka. Note herniation of cyst of the posterior fossa into the upper spinal canal.

made in the first few months of life and wide excision of the cyst membrane carried out with proper supportive measures, the results should be much improved.[33] Such a patient, who was operated on at one and a half months of age, is shown in Figure 141.



Figure 141. Congenital atresia of the foramina of Magendie and Luschka.

A. Pre-operative appearance of hydrocephalic infant at one month of age

B. Same patient at 20 months of age. Development now seems to be progressing at normal rate.

PART III

TRAUMA

Chapter I

Closed Head Injuries

MILD CLOSED HEAD INJURY

A BRIEF SECTION is devoted to the mild closed head injuries of early childhood because they are a matter of such great concern to all pediatricians and general practitioners. Evaluation of minor injuries in this age group is extremely difficult because the nature of early symptoms and signs often makes it impossible to differentiate minor inconsequential bumps from potentially serious trauma.

It is common in infancy for head injury, whether mild or severe, to be accompanied by furious crying, followed by vomiting, pallor, general apathy, lack of responsiveness, and a desire to be left alone. How much of this is due to fright is often difficult to judge. Absence of reliable history of the details of an accident or, indeed, of the occurrence of the accident itself is common. The extreme rapidity of change of a child's vital signs and level of consciousness may be alarming.

All of these factors emphasize the supreme importance in infancy and childhood of what is perhaps the most significant feature in the management of all patients with closed head injuries; namely, repeated careful notation of the vital signs and neurological status at regular intervals until complete recovery has occurred. Obviously this does not mean hospitalization and thorough neurological examination for all of the many mild injuries of early life. However, every youngster who has been rendered unconscious by a blow on the head should certainly be seen by a physician and should be kept under frequent observation until his state of consciousness returns to normal. When children have been only momentarily stunned, they are usually better managed at home under the care of their parents. It is wise under these circumstances to set certain intervals at which parents should observe the child's alertness, motion of all extremities and equality of the pupils. During the first night after injury the child should be awakened two or three times to be certain he is in a natural sleep and not slipping into an unresponsive state. Parents should be cautioned to call the physician for increasingly severe or persistent headache or vomiting, development of extremity weakness, inequality of pupils or increased drowsiness. If the child's appetite and alertness return quickly to normal, treatment should be limited to intelligent observation and gradual increase of activity.

There are few experiences more disturbing to parents than to see their child unconscious from a head injury, particularly when the injury may have resulted through some real or imagined fault of their own. The possibilities of impairment of mental development, of convulsions, of permanent paralysis loom large. As soon as it is justified, therefore, the physician should reassure them of the probable complete recovery without sequelae which they may expect from this type of injury.

SEVERE CLOSED HEAD INJURY

First-Aid

An immediate appraisal of the child's general condition including a survey of the extent of possible injury elsewhere is of prime importance. The establishment and maintenance of an adequate airway then takes precedence over all other diagnostic and therapeutic maneuvers. In comatose children, the cough, gag and swallowing reflexes may be absent or so depressed that normal secretions quickly accumulate in the respiratory tract. Vomiting is particularly frequent after head injury in childhood, so that aspiration of gastric contents is a constant danger.

The comatose child should be placed so that gravity will aid the drainage of nasopharyngeal and oral secretions. Mechanical suction should be made available as soon as possible. Direct laryngoscopy with tracheal aspiration or actual bronchoscopy should be employed if necessary. Occasionally elective tracheotomy may be necessary, to provide adequate oxygenation in profoundly comatose children, particularly when facilities for constant nursing care are limited. Oxygen should be administered during and after these manipulations if there is cyanosis.

Profound shock is rarely seen in closed head injury in the absence of loss of blood or severe injury elsewhere in the body. The usual milder type of shock in childhood responds well to external warmth, rest, relief of pain, reassurance, or perhaps one intravenous infusion of plasma or whole blood. Treatment of shock should, of course, precede all other treatment except maintenance of the airway.

Perhaps the most significant step in management of the child with a severe head injury, once his airway and peripheral circulation have been checked, is early establishment of a base line with respect to certain vital signs. These should be initially recorded as soon as possible and at regular subsequent intervals in order to evaluate progress. They include temperature, blood pressure, and the rate and rhythm of the pulse and respirations. If the vital signs remain constant, conservative observation may be pursued with considerable assurance and the interval of charting may be gradually lengthened. A tendency for the blood pressure to rise steadily or the pulse

or respiratory rate to diminish is much more significant than any single set of observations and usually means increasing intracranial pressure.

Clinical Examination

A brief neurological examination should be carried out as soon as the child's general condition warrants. With uncomplicated cerebral concussion or contusion, maximum neurological deficit usually occurs at the moment of injury and from that point on the child remains fairly stable or steadily improves. When new neurological signs develop during a period of observation after injury, particularly if these are localizing, it usually means that operative intervention is indicated to deal with or rule out intracranial hemorrhage. Elaborate neurological studies are unnecessary. The following observations are the most significant in childhood:

State of Consciousness: If the patient is old enough and sufficiently alert to obey commands, his level of consciousness can be followed by the accuracy and speed with which this is done. If he is too young or too stuporous to respond to questions, the best method of testing consciousness is the type of response to some standard painful stimulus, such as pulling hair or pinching the skin of the neck. Evaluation of swallowing, gagging and eyelid reflexes is also valuable.

Eyes: The size, equality, and response to strong light of the pupils can be determined readily at any age. If both pupils are very small and do not dilate in response to a painful stimulus in a comatose child who has had no medication, the outlook is fairly grave and usually means mid-brain involvement. If both pupils are dilated maximally and fixed to strong light in the acute phase of head injury it probably always signifies a hopeless prognosis, this situation is usually accompanied by respiratory irregularity, fever and decerebrate rigidity, or later, complete flaccidity. If the pupils are unequal, it is indicative of localized injury. When unilateral dilatation of a pupil appears during observation, it usually means intracranial hematoma formation and usually on the side of the dilated pupil. When minor pupillary inequality is present from the moment of injury, it may be associated with contusion alone.

Examination of the eye grounds in infants and children after head injury may be extremely difficult and is important only in visualization of hemorrhage and establishment of the normal appearance of the optic discs.

Motor and Reflex Activity: In infancy, careful observation of spontaneous activity as well as motor response of the extremities to painful stimuli is most important. In older children voluntary motor weakness, hypertonicity and pathological reflexes are easier to evaluate. Motor and reflex responses of both sides should be compared at each examination and from one examination to another. The appearance of unilateral increase in

the deep tendon reflexes or in motor weakness or spasticity during a period of observation is especially indicative of localized intracranial hemorrhage. Extensor rigidity of all extremities, when present early after injury, carries an ominous prognosis.

In summary, it is well to emphasize that repeated regular observation of these few signs is of much greater significance than any single complete neurological study.

X-ray Examination

In the majority of children with closed head injuries, roentgenographic examination contributes little or nothing to the immediate care of the patient after injury and serves principally in ruling out certain types of fracture which necessitate special treatment. It is well to delay x-ray studies, therefore, in restless, overactive, uncooperative children and in those who are vomiting or having any embarrassment of respiration or peripheral circulation. Portable roentgenograms of the skull, particularly in children, are usually worthless. If an acute surgical emergency is suspected, such as an extradural hemorrhage, decision to operate is based on clinical signs rather than x-ray findings.

Lumbar Puncture

Lumbar puncture is not "routine" in the treatment of closed head injuries in childhood in this clinic. There is little indication for its performance in the acute phase of injury and it may be poor judgment to subject a child who is in poor general condition from shock or respiratory embarrassment, or who is simply frightened unduly, to this additional disturbing experience. There should be no hesitation in performing lumbar puncture, however, if it is felt that it may provide any information valuable to the patient's immediate care. Often it is helpful to know the extent of subarachnoid bleeding associated with closed head injury, and a single diagnostic tap can be done at a convenient time when the patient has stabilized.

Lumbar puncture is often of value in children who show persistent headache or restlessness not controllable with ordinary amounts of sedation. Under such circumstances the pressure may be 200 to 400 mms. of water and its gradual reduction to approximately half the initial level is safe and may be helpful symptomatically. There seems to be no point in repeated lumbar punctures in an effort simply to remove small amounts of blood from the subarachnoid space. Lumbar puncture is of no particular value in the diagnosis of subdural hematoma since the lumbar spinal fluid may be entirely clear and under either normal or increased pressure.

Stiffness of the neck and unexplained fever appearing during convalescence are indications for diagnostic lumbar puncture.

Fluid Therapy

Children with head injuries should be adequately hydrated as judged from moisture of mucous membranes and volume and specific gravity of the urine. It is unwise to force fluids, but it is equally unwise to restrict them to a point of dehydration. If an injured child is conscious, he should be allowed to take fluids as desired by mouth. If he is comatose, fluids can be given parenterally early after injury and later by stomach tube. When intake is limited to parenteral fluids, the amount of saline should be restricted to about 10 ccs. per pound of body weight per 24 hours. Bowels should be kept open, but strong purgatives are not indicated.

Intravenous administration of hypertonic solutions is often helpful for temporary reduction of cerebral edema following head trauma. Fifty per cent glucose or sucrose, concentrated plasma, and human albumin have all been used effectively in infants and children. It must be remembered their effect is transient, and therefore they should be repeated at intervals of three to four hours until increased intracranial pressure remains below a critical level.

Sedation

All forms of sedation should be used sparingly following severe closed head injury in childhood. Perhaps the most useful drug is aspirin given in suitable amounts by mouth, stomach tube or rectum. For ordinary headache, restlessness and anxiety this is usually sufficient; if not, the addition of small amounts of codeine, demerol or one of the barbiturates is recommended. For the extremely overactive, unmanageable child, paraldehyde by rectum or intramuscularly is probably the best drug.

If convulsive seizures occur in the acute phase of closed head injury, anticonvulsant medication should be administered at regular intervals, usually in the form of phenobarbital, and continued for some time after clinical recovery depending upon electroencephalographic studies.

Hyperthermia

In childhood, cerebral contusion, with or without intracranial hemorrhage, is particularly prone to produce abnormal elevation of temperature in spite of adequate hydration and absence of infection. Persistent extreme hyperthermia in the acute phase of head injury carries an ominous prognosis, but by no means always signifies irreversible damage and should, therefore, be treated vigorously. Artificial means of controlling body temperature include: administration of large amounts of salicylates, application of ice bags, alcohol sponging of the exposed body surface, the use of an electric fan, and ice water enemas. In infancy and early childhood the

temperature may change very abruptly, so it should be checked frequently during treatment to prevent too rapid cooling.

Chemotherapy

All comatose patients who are not taking care of oral and nasopharyngeal secretions properly, and those on constant bladder drainage should receive prophylactic chemotherapy. Penicillin, 300,000 U daily is usually adequate. If x-ray films demonstrate a basal skull fracture or a fracture extending into the mastoid or paranasal air sinuses, prophylactic chemotherapy is also indicated.

Ambulation

Early ambulation and rehabilitation of children with closed head injuries is just as desirable as it is in adult patients, and usually much less of a problem. As soon as vital signs have remained stable for a reasonable period, if the neurological examination is negative and the child free from severe headache and nausea, mobilization should be encouraged. It is neither necessary nor desirable for children with cerebral concussion and contusion, *even in the presence of linear fracture of the skull or mild subarachnoid* bleeding, to remain flat in bed for long periods providing their clinical progress is satisfactory. The amount of activity should be increased gradually rather than abruptly. Extremes of physical fatigue, over-eating and constipation should be avoided. There seems to be no doubt that the frequency of extended convalescence and disturbing post-traumatic symptoms can be diminished markedly by intelligent early ambulation.

The incidence of various types of head injury in children under 12 years of age admitted to the neurosurgical service of The Children's Medical Center for treatment is shown in Table IX. Many additional children, of course, are seen daily in the emergency ward with concussion and even linear skull fractures and returned home after a period of observation.

TABLE IX

CHILDREN HOSPITALIZED BECAUSE OF HEAD INJURY
1939 to 1951

Cerebral Concussion and Head Injury, unclassified	374
Cerebral Contusion	89
Cephalhematoma	28
Skull Fracture, simple	285
Skull Fracture, depressed	146
Skull Fracture, compound	59
Extradural Hematoma	30
Subdural Hematoma	319
Total	1,330

Chapter 11

Fractures of the Skull

Linear Fracture

UNDER ORDINARY circumstances linear fracture of the calvarium in infancy and childhood does not in itself alter the management from that of closed head injury as set forth in the previous chapter. In early childhood, linear skull fracture without separation usually is healed so that it can no longer be detected on x-ray examination within three to four months. No special head protection is indicated. Fractures which cross the pathway of the middle meningeal vessels or the lambdoid suture should always alert the observer to the possibility of extradural hemorrhage.

Linear fractures, particularly those of the parietal region in infancy, are often associated with fairly extensive subgaleal hematomas. Palpation of the edge of these hematomas gives the impression of an actual bony depression, and frequently this possibility can be resolved only by x-ray examination. The treatment of these subgaleal hematomas and of the cephalhematomas associated with difficult delivery is almost always conservative. Unless there is evidence of continued bleeding with the clot enlarging so that the scalp becomes stretched and tense (Figure 142), aspiration is not indicated; usually the clot will be absorbed within a few days to weeks. If aspiration is performed it should be done through a widely shaved and aseptically prepared area of the scalp using a needle fitted with a stilette. Contamination of a cephalhematoma in infancy may result in a very indolent, serious infection extremely difficult to treat (Figure 143). Following aspiration, a light circular pressure dressing will help prevent reaccumulation. Large cephalhematomas occasionally calcify (Figure 144). There is no indication for surgical treatment unless these are grossly disfiguring.

Diastatic Fracture

In early childhood, traumatic separation of the cranial bones at the suture lines occasionally occurs. Presumably fibrous tissue is torn and the bones spring apart rather than being fractured themselves. These "diastatic" fractures most commonly involve the lambdoid suture (Figure 161) and

in a number of cases have been associated with extradural hematomas due to rupture of emissary veins or posterior branches of the meningeal vessels. No special treatment is indicated because of the suture separation itself. The child should be managed as a closed head injury, but observed very carefully for evidence of intracranial bleeding.

Basal Skull Fracture

Fracture through the basilar portion of the skull is not common in childhood but when it does occur is treated as in adults. If there is bleeding into the nose, nasopharynx, middle ear, or mastoid after head injury it is wise to assume that a basilar fracture is present even though it may not be possible to demonstrate it by x-ray examination. Cerebrospinal fluid rhinorrhea, otorrhea, or pneumocephaly (Figure 145) implies a basilar skull fracture. The same is true of a peripheral type facial nerve paralysis since these usually occur with fracture through the petrous or mastoid portions of the temporal bone. Fractures of the sphenoid bone or the base of the frontal fossa are usually accompanied by hemorrhage into the orbit and periorbital tissues (Figure 146).

It is wise to treat all basilar skull fractures as compound lesions as far as chemotherapy is concerned. Prophylactic penicillin for seven to 10 days at least, and longer in the presence of fever, meningeal signs or spinal fluid leak is advisable. Aside from prophylaxis against infection, however, treatment is essentially that of closed head injury and is determined by the progress of the vital and neurological signs.



Figure 142. Eighteen months old infant with huge subcutaneous hematoma resulting from a fall on the forehead Patient has hemophilia. Liquid blood and clot were evacuated after patient's clotting time had been returned to normal by fresh plasma infusions.



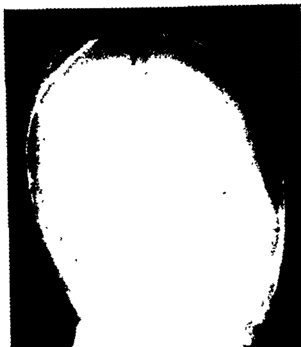


Figure 143 (*Left*). Characteristic indolent, chronic burrowing infection of the scalp due to *b. coli* contamination of a large cephalhematoma dating from birth

Figure 144 (*Right*). Calcification in a subgaleal hematoma of the left parietal region resulting in palpable and visible deformity.





Figure 145 Antero-posterior and lateral roentgenograms of 7 3/12 years old boy three weeks after head injury Patient had rhinorrhea at this time. Films show large collection of air in the intracranial chamber associated with a long comminuted fracture of the skull which extended into the cribriform plate.



Figure 146. Appearance of three and a half year old boy following severe frontal injury with fracture of the sphenoid bone and cribriform plate with rhinorrhea. Patient also had transient blindness and diabetes insipidus. Complete recovery.

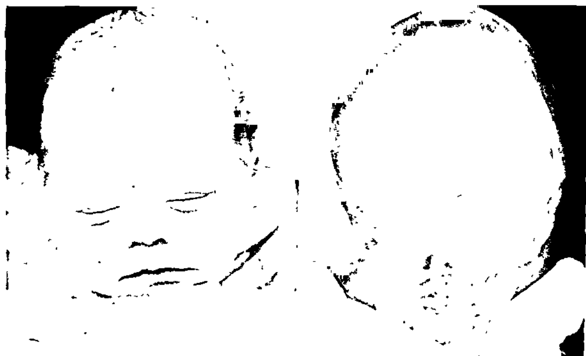


Figure 147 (*Left*). New-born infant with depressed fracture of the right frontal bone incident to delivery. Negative neurological examination and uneventful course following elevation of the fracture.

Figure 148 (*Right*). Antero-posterior roentgenogram of new-born infant showing depressed fracture of the left parietal bone. Note that this produces some deformity of the sagittal suture line as well

Depressed Fracture

Because of the rapid growth of the brain during the first two years of life it is particularly important in this age group that all depressed fractures of the skull be elevated as soon as possible. Localized external compression of the brain may serve not only to interrupt function but may also interfere with growth and perhaps establish an epileptogenic focus.

Depressed fractures are commonly seen in the new-born period as a result of extreme molding of the head during its passage through the birth canal or as a result of forceps application or digital pressure by the obstetrician at the time of delivery (Figure 147). Such depressions vary from a small dent not more than a centimeter or two across to marked depression of virtually an entire frontal or parietal bone (Figure 148). Bilateral parietal depressed fractures may occur secondary to difficult forceps extractions (Figure 149). These injuries are comparable to "green stick" fracture of the long bones in childhood. That is, there is often no actual break in continuity but the bone is bent inward and stays in this depressed position.

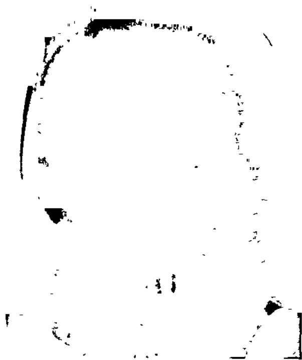


Figure 149. Bilateral depressed fracture of the skull in a new-born infant secondary to difficult forceps delivery.

Treatment consists in surgical elevation of the depressed bone; these fractures do not reduce themselves spontaneously. Reduction should be performed as soon as the infant's general condition is stable. New-born infants tolerate this type of surgery extremely well within the first day or two of life.

A short linear or curved scalp incision is made along one margin of the depressed area. A small burr hole is drilled in normal bone and a narrow blunt instrument such as a periosteal elevator introduced under the depressed bone which is then gradually levered back into position (Figure 150). Occasionally it "pops" back in one quick motion, but usually it is necessary to maneuver it back more slowly by changing the position of the elevating instrument several times. If the depressed bone is impacted so tightly that it will not move with reasonable pressure, or if it can be elevated

but not held in normal position, it is wise to make a linear cut in the bone with stout scissors from the burr hole to the center of the area of depression. This will allow the fragments to be elevated usually without difficulty.

These operations are carried out under local anesthesia usually supplemented by a sugar nipple. The latter can be reinforced by a few drops of paregoric or brandy to good advantage. Prematurity is no contra-indication to operation as long as the baby's temperature is controlled.

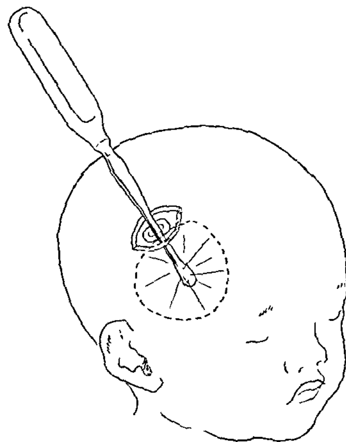


Figure 150. Method of elevating depressed fracture of the skull in infancy. Periosteal elevator is inserted through burr hole exposed by small scalp incision to lever depressed bone back into normal position.

Depressed skull fractures in older infants and children are treated essentially as in adults. If the injuries are not compound or extremely comminuted, the bone fragments are preserved.

Compound Fracture

Compound cranio-cerebral trauma occurs rarely in infancy but fairly commonly in childhood. Vehicular accidents are the commonest cause. In addition, youngsters are often struck by flying rocks, base ball bats and golf clubs, and their heads seem prone to strike sharp objects in falls from trees, porches and fire-escapes.

First-aid treatment should consist in shaving and cleaning the scalp widely around the wound followed by application of a sterile dressing and institution of chemotherapy, preferably penicillin. There is no preliminary first aid-surgery appropriate to compound cranio-cerebral injuries. The initial surgery should be definitive. Since this may mean a complicated and prolonged operative procedure involving all layers of the scalp, skull, meninges and brain itself, it should be delayed until the proper personnel and facilities are available. With antibiotic therapy such delay is entirely safe.

Children should be out of shock and usually under endotracheal inhalation anesthesia before surgical treatment is attempted. Whole blood for transfusion should be available. The area is widely prepared with soap and

water and mild skin antiseptics. The scalp is first debrided by removal of all devitalized and grossly contaminated tissues. In the debridement of bone it is unwise to attempt to preserve fragments which have been grossly contaminated or badly comminuted. The brain wound itself is debrided by careful use of gentle suction with accurate hemostasis, always if possible under direct vision. All foreign bodies and bone fragments should be removed from the brain. The brain wound should be left clean and dry as in any elective surgical procedure.

Defects in the dura are sutured tightly, grafted with periosteum or replaced with polyethylene *film*. In occasional clean compound wounds treated early, either replacement of loose bone fragments or primary cranioplasty will be possible. In general, however, repair of skull defects should be deferred. It may be necessary to perform primary or secondary plastic revisions of the scalp in avulsed wounds before cranioplasty can be carried out (Figures 151, 152 and 153).



Figure 151. Defect remaining after suture of avulsion injury of scalp associated with underlying compound depressed fracture of the skull.

Figure 152 Appearance of scalp after shifting large pedicle flap to cover defect caused by avulsion injury shown in Figure 151.

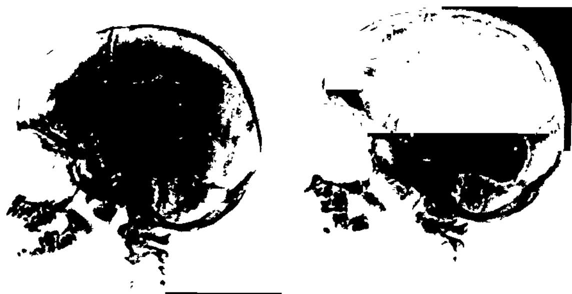


Figure 153 Lateral roentgenograms of patient shown in Figure 151 and Figure 152 before and after tantalum cranioplasty.

Chapter III

Cranioplasty

In infancy and early childhood there is little need for repair of traumatic skull defects, particularly if the underlying dura is intact, because new bone forms quickly at this age from periosteum and dura. In older children, however, the indications for cranioplasty are essentially the same as in adults. Skull defects above the insertion of the temporal and occipital muscles which are larger than 2 to 3 cms. in diameter should in general be closed. If these defects are in the forehead, the visible deformity alone is indication for cranioplasty. Other reasons for repair are the vulnerability to direct injury of uncovered areas, the occurrence of unpleasant symptoms such as local throbbing when the head is lowered, and the tendency for

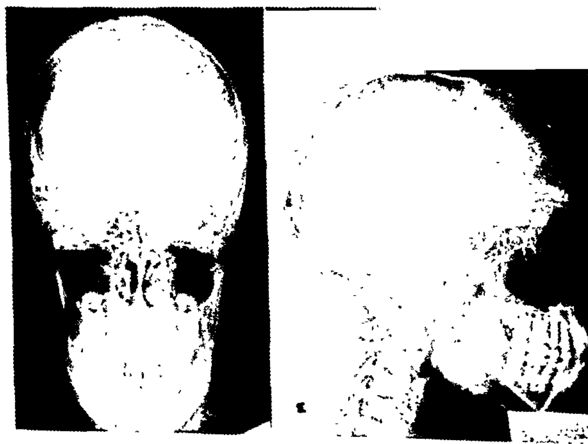


Figure 154 Postero-anterior and lateral roentgenograms of the skull of a 14 year old boy following debridement of a severe compound, comminuted, depressed fracture of both frontal bones with laceration of both frontal lobes of the brain due to penetration of a sky rocket.

ventricular enlargement or "migration" toward a skull defect, particularly if the underlying dura is also deficient.

In childhood four types of cases have been encountered in which cranioplasty has been indicated.

(1) By far the largest number of patients have been those with skull defects secondary to compound fracture of the skull, as considered in the previous section (Figures 154 and 155). Occasionally primary cranioplasty at the time of debridement of the fracture can be performed, but usually it is delayed from a few weeks to a few months after healing of the original wound. Delayed leptomeningeal cyst formation due to unrecognized dural laceration associated with closed linear fracture may also necessitate cranioplasty (Figures 156 and 157).

(2) The second group of patients include those with congenital defects of the skull.[150] There are many small congenital defects which fill in partially or completely as growth proceeds. However, in some patients with simple absence of a large area of bone or with a large defect associated with a meningocele of the cranium, bone never regenerates satisfactorily and cranioplasty becomes necessary (Figures 102 and 107).

(3) The third group of patients constitutes the children with severe

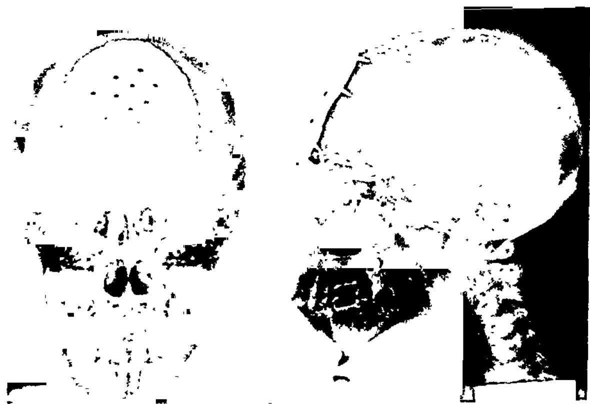


Figure 155. Antero-posterior and lateral roentgenogram of the same patient as in Figure 154 following tantalum cranioplasty. Note that an adequate bridge of bone was left in the supra-orbital region so that tantalum plate could be firmly inserted. Good cosmetic result. Asymptomatic two years after cranioplasty.



Figure 156. Postero-anterior and lateral roentgenograms of the skull of a five year old child showing parietal bony defect on the right due to leptomeningeal cyst. Patient had head injury two years previously.



Figure 157. Postero-anterior and lateral roentgenograms in the same patient as Figure 156 following closure of the cranial defect by tantalum cranioplasty.

lead encephalopathy in whom large bone flaps have been removed in an effort to treat extreme increased intracranial pressure seen in the acute phase of this disease (p. 424).

(4) The fourth group includes those in whom bone has been sacrificed at operation because of tumor or infection. These include primary tumors of the skull (Figure 289), tumors of the meninges (Figure 294), osteomyelitis, and post-operative infection or aseptic necrosis.

After three to five years of age there is relatively little growth of the skull, and there should be no hesitation beyond this period in performing cranioplasty for any of the foregoing indications.

In this clinic, three materials have been used for cranioplasty in children; bone, methyl methacrylate (Leucite), and tantalum. Bone is particularly appropriate for repair of congenital defects in the supranasal and supraorbital areas. The squamous portion of one or both temporal bones may be taken as a free graft from beneath the temporal musculature (Figure 49). Thin strips of rib may likewise be used.

Methyl methacrylate plates are light, radio-lucent and inexpensive. They require very careful molding, however, in a two-stage operative procedure. They are brittle, and the cosmetic results have not been satisfactory in growing children.

By far the most widely used material in this clinic has been tantalum. Tantalum plates which have been carefully made, accurately fitted, and securely inserted have given the best cosmetic and functional results in children from three years of age up. With present techniques, this has also been the most convenient method. A one-stage procedure is employed. A cast-iron positive and negative skull mold is used on which to fashion the plate (Figure 158). The plate is fashioned roughly on these molds prior to operation and then finished during operation, at which time the cast-iron molds are sterilized and available to the surgeon in the operating room. All of our plates are countersunk in a ledge made by removing outer table the thickness of the tantalum plate, and are held in place by tantalum glaziers' points.[85]

The most frequent complication of tantalum cranioplasty in childhood has been denting of large plates by falls against blunt objects. In general, the cosmetic and functional results have been excellent. Since using the cast-iron sterile skull model, 33 tantalum plates have been inserted in children between three and 12 years of age. *Of these, one was removed and immediately replaced one week after insertion because of a poor fit, and one was removed after five months because of a chronically infected granuloma beneath the plate. This latter was replaced successfully six months later. All of the other results have been entirely satisfactory.*

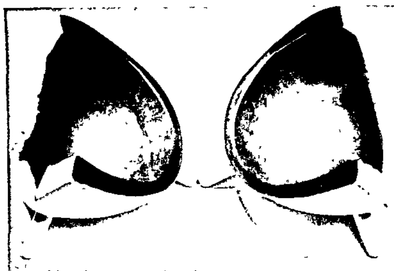


Figure 158A. Positive and negative cast-iron models of a small female adult skull used for fashioning tantalum plates These models are sterilized and facilitate accurate fashioning of plates at the operative table.

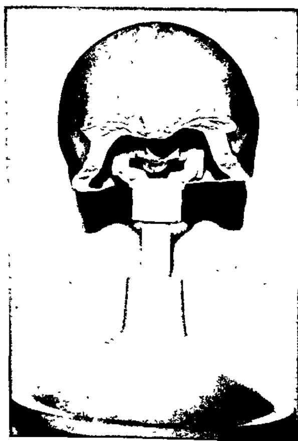


Figure 158B.

Chapter IV

Extradural Hematoma

EXTRADURAL HEMATOMA, though less common in infancy and childhood than among adult patients, constitutes one of the most critical and urgent complications of closed head injury in the pediatric age group. Unless this condition is recognized early and surgical therapy instituted promptly, the mortality rate is high.

Whereas in adults extradural hematoma is almost always due to arterial hemorrhage from middle meningeal vessels which have been lacerated by fractures through the squamous portion of the temporal bone, in infancy, *extradural bleeding may also occur from dural veins and from torn emissary* veins in the region of the major dural sinuses. In children, extradural bleeding may occur in the absence of demonstrable fracture of the skull, or with simple diastatic fracture, particularly of the lambdoid suture. If extradural bleeding is from the middle meningeal artery, the clinical course in infancy may be extremely rapid, but if bleeding is largely venous, symptoms and signs may be delayed for many hours or even days after injury.

It has generally been felt that extradural hematoma in early life is extremely rare. However, its frequency in this clinic [28] (Table IX) suggests it is common enough to warrant detailed consideration in this text. Thirty patients under 12 years of age with massive extradural hematomas provide the basis for this discussion. The age distribution of these patients is shown in Figure 159. It will be noted that more than half occurred in the first two years of life. Twenty-one of the patients were male and nine were female.

Symptoms and Signs

In adults the classical history of extradural hemorrhage is initial loss of consciousness at the time of head injury followed by a lucid interval and then subsequent lapse into coma. In childhood, the initial period of unconsciousness occurs rather rarely. It is more common for a child to be momentarily stunned, then cry vigorously, and subsequently, after a variable latent period, develop more and more drowsiness which may progress slowly to stupor, or occasionally with great rapidity to profound coma. As stressed in the section on closed head injury, whenever the level of consciousness deepens, or localizing neurological signs appear during a period of

observation after injury, intracranial hemorrhage must be assumed until proved otherwise.

The commoner clinical features as noted in the cases from this clinic are shown in Figure 160. There is usually local evidence of head trauma; scalp swelling, contusion, laceration or ecchymosis. A visible scalp lesion usually overlies the site of the extradural hematoma. Aside from the development of stupor, the most significant feature in the course of this lesion is the appearance of lateralizing neurological signs. These consist of paralysis of the extremities and the face contralateral to the hematoma and dilatation of the pupil on the same side as the lesion. The development of hyperactive deep tendon reflexes and an extensor plantar response unilaterally may precede objective evidence of weakness.

Because of the rapid increase in intracranial pressure which results from massive supratentorial arterial bleeding, medullary compression may occur early and be manifested by strabismus, respiratory irregularity and depression, bradycardia, and decerebrate postural patterns. In infants, the characteristic clinical picture of intracranial hypertension may be modified considerably, however, by blood loss. The amount of bleeding into

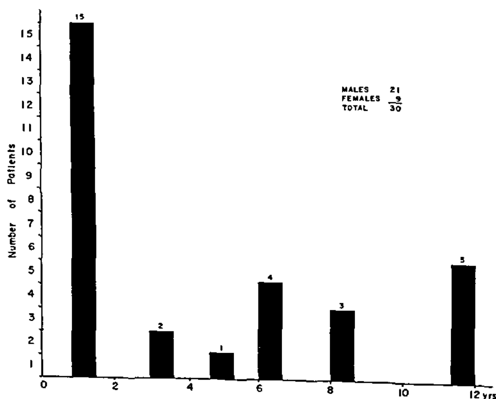


Figure 159. Age and sex distribution in 30 children with extradural hematoma.

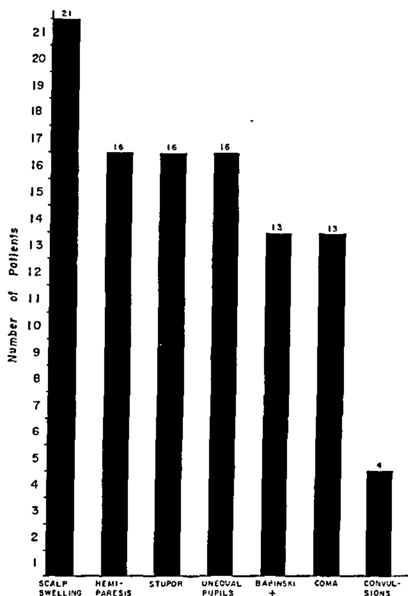


Figure 160 Summary of the common signs elicited in 30 children with extradural hematoma.

the extradural space may be sufficient to produce a rapid and severe anemia and signs of profound shock. Thus, the pulse instead of being full and slow is more apt to be weak and rapid. Blood pressure is low or unobtainable instead of being elevated. The baby is cold, pale and clammy. Obviously, the presence of normal optic discs does not rule out extradural hemorrhage since there usually will not have been sufficient time for the development of papilledema. Lumbar puncture is of little significance in this lesion and should not be performed when the clinical course is one of rapid increase in the severity of neurological or vital signs.

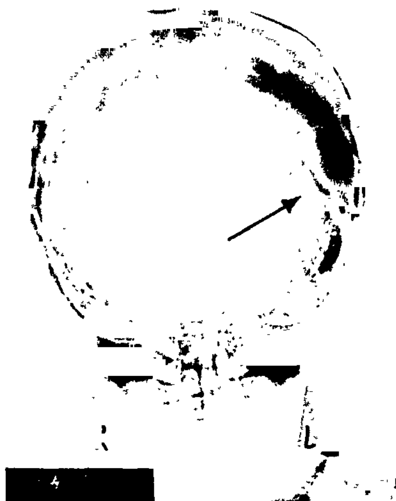


Figure 161. Basal roentgenogram of the skull showing diastatic fracture of the lambdoid suture on the left. This is the type of fracture often associated with tearing of emissary veins and extradural venous bleeding.

X-ray examination should likewise be omitted when the patient's condition demands prompt shock therapy and early surgical exploration. Roentgenograms which demonstrate a linear or depressed fracture crossing the path of the middle meningeal vessels or demonstrate a diastasis of the lambdoid (Figure 161) or squamosal suture should always alert the observer to the possibility of extradural hemorrhage. Figure 162 demonstrates the incidence of skull fracture among the patients in our series.

Operation

Indications for exploration have already been stated, but are emphasized here once more. Any infant or child with a history of head injury, whether or not there are local or x-ray signs of trauma, who develops during observation an increasing degree of drowsiness or stupor, vital signs suggestive of increasing intracranial pressure, or lateralizing neurological signs

(pupils, reflexes, motor activity) should have exploratory burr holes performed.

Usually in this condition, surgery is a real emergency. The child who arrives in the hospital stuporous or perhaps even completely comatose, with impaired respirations, a poor pulse, a unilateral dilated pupil and contralateral hemiplegia should be taken immediately to the operating room and prepared for operation while blood is being cross-matched and constant intravenous infusion started. It has occasionally been necessary to cut-down on a vein in the emergency ward and start plasma or blood before transporting the infant to the operating room. Operative exploration for extradural hematoma should never be commenced until an intravenous infusion has been started and blood for transfusion is available. This is true because release of impacted bone fragments and blood clot at opera-

INCIDENCE OF SKULL FRACTURE AND SUTURE DIASTASIS

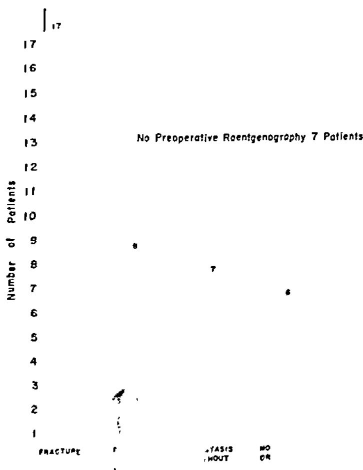


Figure 162. Types of found
oma.

tion may permit resumption of furious arterial bleeding with substantial additional blood loss before the actual bleeding point can be located and controlled.

Exploration in children is carried out under ether anesthesia unless the child is moribund, when local procaine alone is sufficient. After the head is shaved and prepared, a burr hole is placed low in the temporal region over the course of the meningeal vessels. The head should be draped so that incision can be carried down to the zygoma and high up in the parietal region if necessary. A fracture line is frequently disclosed as soon as the scalp incision is made when the diagnosis of extradural clot is confirmed through the burr hole the latter is quickly enlarged to a decompression of sufficient size to allow rapid removal of the clot, usually by suction (Figure 163). This additional bony opening should be carried downward to expose the meningeal vessels as low as possible. Usually the actual bleeding point is readily located and can be secured with a tantalum clip, silk ligature or electrocoagulation. If there is diffuse bleeding from the dura all along the course of the meningeal vessels, it may be necessary to ligate these vessels as low as possible primarily or to plug the foramen spinosum.

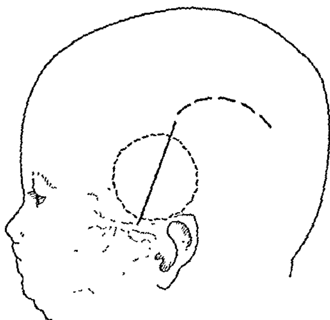


Figure 163. Plan of exposure for evacuation of extradural hematoma in infants and young children. Heavy solid line indicates position of the skin incision with a posterior curve extension if necessary. Dotted line indicates the average extent of bony removal necessary to evacuate the clot and secure the bleeding points.

Extradural hemorrhage of venous origin, as from one of the large emissary veins in the region of the sigmoid sinus, can be controlled usually with the cautery and gelatin foam. Every effort should be made to find the actual bleeding point as quickly as possible and secure it directly (Figure 164). It may be necessary to extend the scalp incision posteriorly (Figure 163) and elevate a scalp flap to gain sufficient exposure in this variety of hemorrhage. The dura may be depressed away from the skull as much as 2 or 3 cms. over a wide area. It usually re-expands quickly when the clot is evacuated. It is wise to make a tiny incision in the dura to explore the subdural space for hemorrhage and inspect the surface of the brain.

If complete hemostasis has not been achieved or the brain does not quickly expand to obliterate the extradural dead space, a drain should be left to this area for 24 to 48 hours. If the patient's condition warrants, a burr hole should also be made on the opposite side to rule out the presence of bilateral hemorrhage. This can be delayed if necessary.

Occasionally, post-operative response following evacuation of an extradural hematoma in a child is dramatic. Usually there is immediate improvement in the hemiparesis and the inequality of the pupils. If the child is *comatose at the time of operation, however, there is more apt to be slower* recovery over a period of several days. Post-operative lumbar punctures are judiciously employed for persistent increased intracranial pressure. Gavage feedings, post-operative transfusions and other supportive therapy are employed as necessary.

Results

The mortality in patients with extradural hematoma in infancy and childhood is less than in adults. Among our 30 patients there were three deaths. One patient died seven hours after operation and 13 hours after

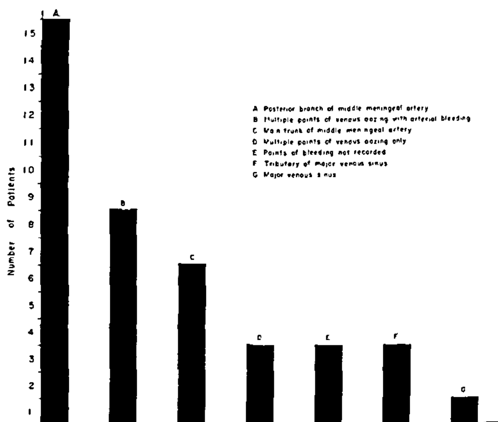


Figure 164. Source of the bleeding as determined at operation in 30 children with extradural hematoma

injury. A second patient died 16 hours after operation and 88 hours after injury; this child had serious brain lacerations as well. The third death occurred in a patient who was moribund and on artificial respiration at the time of operation and who never started to breathe again spontaneously. The recovery in all of the other patients, though occasionally quite prolonged, was excellent, even in extremely critical situations. At least two patients have mild residual hemiparesis. Convulsions post-operatively have been rare.

the birth canal or in the actual events of delivery. It may occur as a result of the moving head striking a solid object, as in falls, where there is a differential rate of deceleration of the skull and the less dense intracranial contents. This type of trauma presumably causes tearing of the delicate veins which bridge from the cerebral cortex to the large dural venous sinuses, principally at the vertex. It should be emphasized, however, that due to frequent inadequacy of the history, absence of a record of head injury should never rule out in itself the diagnosis of subdural hematoma.

Although any disease which tends to increase bleeding in early life probably contributes to the incidence and severity of subdural hemorrhage, the great majority of patients in this clinic have demonstrated no evidence of a bleeding tendency.

Symptoms and Signs

There is no clinical picture characteristic of this lesion. In Figure 167 is shown the frequency with which various symptoms have been noted. It is apparent that the commoner ones, convulsions, vomiting, irritability, and failure to gain weight, are found in many types of disease in infancy, both intracranial and otherwise. Convulsions are of all varieties and all degrees of severity and have been of no lateralizing value.

In Figure 168 is shown the frequency with which the commoner physical findings have been noted. Perhaps commoner than any of these actually is a finding difficult to define; namely, a restless hyperirritability and resistance to being handled or examined. Temperature abnormality is usually due to associated dehydration or systemic infection. Most of these youngsters show diffusely hyperactive reflexes, intermittently or constantly rigid extremities, and unsustained ankle clonus. Attempts to lateralize clot formation on the basis of neurological signs are usually unsatisfactory in infancy.

Slight enlargement of the head with bulging of the anterior fontanelle, separation of the sutures, and dilatation of the scalp veins may lead to an erroneous diagnosis of hydrocephalus (Figure 169). Papilledema is rarely seen, but retinal hemorrhages are common. A frequent clinical picture in the first few months of life is that of an infant who has simply not been doing well as evidenced by failure to gain weight, refusal of feedings, hyperirritability, and intermittent low-grade fever, together with a slightly accelerated rate of head growth and perhaps a history of difficult labor or delivery.

Laboratory studies are of little value except that many of these patients are anemic. X-ray examination of the skull is only occasionally abnormal and is never in itself diagnostic. It may show skull fracture or slight separation of the sutures if the lesion is a chronic one. Pneumoencephalography and ventriculography are unnecessary and undesirable.

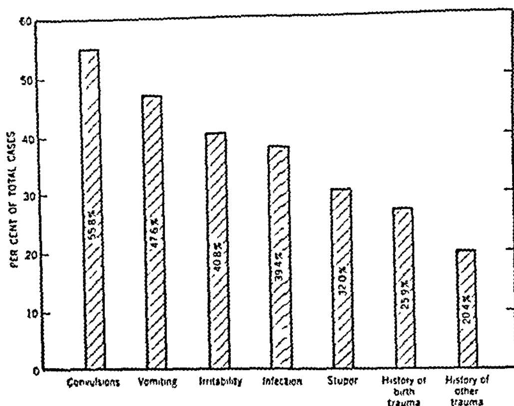


Figure 167. Common complaints elicited in the history from parents of infants with subdural hematoma. (Reprinted through the courtesy of The Interscience Publishers Inc. from *Advances Pediat.*, 4:231-263, 1949.)

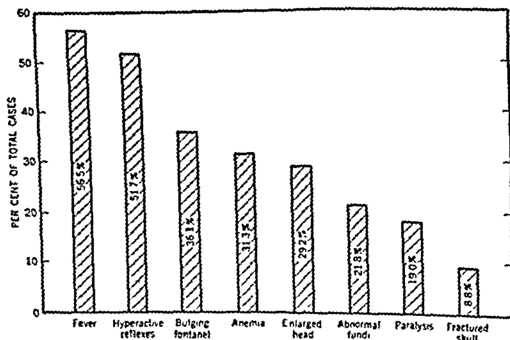


Figure 168 Common physical and laboratory findings in infants with subdural hematoma at time of hospital admission. (Reprinted through the courtesy of The Interscience Publishers Inc. from *Advances Pediat.*, 4:231-263, 1949.)



Figure 169 Ten months old infant with bilateral subdural hematomas. Note shiny scalp, dilated veins, poor nutrition, irritability.

Pathology

In infancy, bleeding into the subdural space probably occurs in almost every instance from ruptured bridging veins along the longitudinal sinus in the fronto-parietal region. As a result, the distribution of clots has been practically uniform. They have extended from the mid-line well over the frontal pole and down toward the floor of the frontal and middle fossae inferiorly but seldom as far posteriorly as the occipital region. Many are limited to the frontal region entirely. In early life, subdural bleeding is usually bilateral (85 per cent), though often more severe on one side than the other.

The pathological characteristics of the hematoma vary with the age of the lesion. In the acute phase of hemorrhage the fluid obtained from the subdural space may be bright red. Subsequently it becomes a deeper reddish-brown or chocolate color, and in chronic lesions it shows varying degrees of xanthochromia with progressively fewer red cells in the sediment (Figure 170). It may occasionally be clear and almost colorless but have a markedly elevated total protein content. Solid hematoma may be



Figure 170. Progressive change in character of fluid removed from the subdural space. Red cells and evidence of hemolysis diminish and xanthochromia of supernatant fluid increases and then becomes less.

present in any stage from bright red fresh clot to old greenish-gray masses of fibrin.

Within a week to ten days after subdural bleeding a membrane begins to form from the inner surface of the dura. In a well developed clot this outer membrane is from .5 to 2 or 3 mms. thick and is a tough, inelastic, shiny gray or reddish-gray vascularized structure (Figure 171). An inner membrane between the clot and the pia-arachnoid also forms eventually. This is usually thin and often quite transparent but may also show well developed blood vessels in chronic lesions (Figure 172). The degree of maturation of these membranes as well as the character of the subdural fluid depends on how early removal of the fluid has been started.

In chronic, neglected subdural hematoma in infancy, the cerebral cortex may show marked atrophy. The convolutions become small, the sulci deepened, and the lateral ventricles dilated. The arachnoid appears gray, thickened and translucent, and there is an excess of subarachnoid fluid. There is marked discrepancy between the size of the brain and the size of the cranial vault (Figure 173).

Treatment

In the first years of life, while the brain is growing rapidly in volume, it is essential to avoid restriction of this expansion if impairment of mental development is to be prevented. It is important, therefore, not only to remove the fluid and solid clot which collects in the subdural space but also to excise the hematoma membranes which form from the under surface of the dura. These membranes, even though very thin, usually prevent complete drainage of subdural hematoma fluid, and in addition they constitute inelastic, connective tissue structures which do not grow with attempted expansion of the brain; as such, they may interfere with development of cortical function.

Immediate evacuation of large subdural hematomas in infancy carries

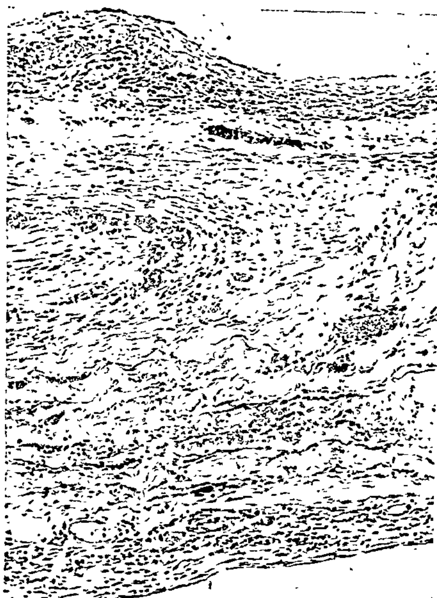


Figure 171. TBE x 200 Photomicrograph through outer membrane of a subdural hematoma removed from a 1 7/12 year old infant. Note the longitudinal arrangement of dense fibrous tissue with numerous well developed blood vessels. There is a mesothelial membranous surface on both aspects with compact arrangement of the fibrous tissue immediately beneath it.

a high mortality. An early, but gradual release of pressure followed by eventual radical excision of membranes when the infant's general condition has improved is preferable. The essential steps in this regimen of treatment consist in (1) subdural taps for gradual removal of fluid until active bleeding has ceased and the infant's condition improved; (2) exploratory burr holes to establish whether or not a membrane has formed; and (3) craniotomy with wide excision of hematoma membranes and evacuation of all solid clot. These steps will now be outlined in detail.

Subdural Taps: The subdural puncture which establishes diagnosis also

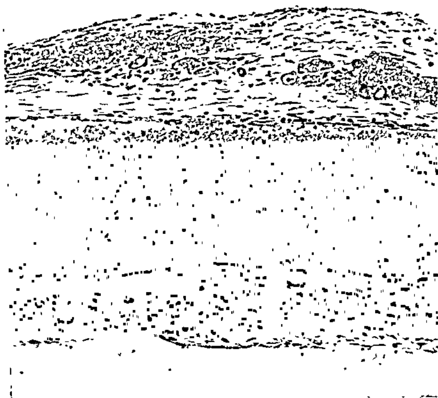


Figure 172. TBE x 200 Photomicrograph of section through inner membrane of subdural hematoma removed from the same patient as Figure 171. This membrane is thinner with a much looser arrangement of the fibrous tissue, and a large and many small spaces containing blood are present.

marks the beginning of treatment. Initially punctures are carried out on both sides. When positive, 10 to 15 ccs. of fluid only are removed from each side. It is unwise to remove more than this, particularly in very ill infants, and an attempt to drain off all the fluid available bilaterally may even be fatal. Subsequent taps are carried out daily on alternate sides. If the fontanelle becomes tight again quickly, or the baby's clinical behavior is poor, it may be necessary to tap oftener than this, but more than 15 to 20 ccs. should still not be removed at any one puncture. A lumbar puncture should be performed at the time of the diagnostic subdural taps to establish the possibility of associated subarachnoid bleeding.

It is wise to keep a sample of each subdural tap so that the character of the fluid may be followed. Usually the number of red cells diminishes and the total protein of the supernatant fluid increases (Figure 170). The infant's condition usually improves rapidly as these taps are continued. He stops convulsing and vomiting, begins to eat well and gain weight and becomes more attentive and less irritable. Transfusions and parenteral vitamins are given to supplement the diet and infection is treated with antibiotics. Usually, after three to 10 days on this regimen, all evidence of

active bleeding has ceased and the baby's condition warrants more radical exploration.

The technique recommended for subdural puncture is as follows. It is a simple procedure and a safe one if carried out with meticulous asepsis and attention to detail. The equipment necessary is minimal (Figure 174).



Figure 173. Extreme cerebral atrophy in a three month old infant with huge bilateral subdural hematomas. Illustration shows the deep space remaining after removal of thick well-organized outer and inner membranes and evacuation of a large fluid hematoma. Prognosis for mental development is extremely poor.



Figure 174 Equipment used for subdural puncture in infants. This includes sterile gloves, sterile towels, skin preparation solution, hypodermic syringe and needle for novocaine injection, No 19 short bevel, two-inch lumbar puncture needles.

These punctures are performed through the coronal suture well lateral to the margin of the anterior fontanelle; the point selected should always be at least one and one-half inches from the mid-line (Figure 175).

The baby is wrapped in a restraining sheet and given a sugar nipple to suck. All of the hair is shaved anterior to a line between the ears and this entire area of scalp is prepared and draped as carefully as for craniotomy. A novocaine wheal is raised and a No. 19 or No. 20 short-bevelled, two-inch lumbar puncture needle with the stilette in place is introduced at right angles to the surface. The shaft of the needle should be grasped and guided



Figure 175 Subdural tap Note position of the needle well lateral to the mid-line The tap is done under local anesthesia with the baby carefully restrained. A sugar nipple is used for analgesia. *The shaft of the needle is held firmly at the surface of the scalp at all times.*

with the thumb and forefinger of the left hand at all times to prevent lateral deviation of the point and to control the rate of penetration accurately. The suture line is easily located by moving the point of the needle back and forth slowly after the scalp has been penetrated. Entry into the subdural space is readily detected by the sudden decrease in resistance felt when the dura is perforated.

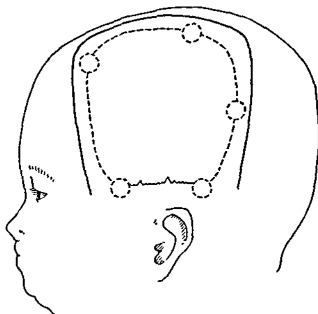


Figure 176. Plan of exposure for removal of subdural hematoma membranes in infants Heavy line indicates the skin incision. Dotted lines indicate the extent of the bone flap The superior-anterior burr hole is first made to establish the existence of a membrane before the rest of the skin incision is made.

Ordinarily only a few drops of clear fluid can be obtained in this manner from the subdural and subarachnoid space. If there is an abnormal amount of subdural fluid, it will flow through the needle as soon as the dura is penetrated. There is no virtue, and there may be considerable danger, in introducing the needle further if fluid is not obtained immediately beneath the dura. Subdural fluid should be allowed to drip from the needle at its own rate and should not be aspirated with a syringe.

A sterile dressing is left over the puncture site between taps. It is wise to avoid repeated taps at exactly the same point in order to prevent spontaneous leakage.

Burr Holes: For the reasons outlined previously, it is desirable that every infant who has had excessive bloody or xanthochromic fluid recovered from the subdural space be explored for the possibility of membrane formation. This is accomplished through small burr holes, which may be performed bilaterally in the parieto-temporal region as a separate procedure or may be placed initially on one side as part of a bone flap which is then completed if the burr hole exploration is positive. In general, if there have been large amounts of xanthochrómic fluid repeatedly obtained on subdural taps, it is best to outline a bone flap, then make the superior, anterior burr hole to confirm the existence of a membrane before craniotomy is performed (Figure 176). The same program is then carried out on the opposite side a week later.

If the amounts of fluid obtained have been small and decreasing rapidly in amount, if the subdural taps have been started in the acute phase after a known head injury, or if the baby's condition is extremely precarious, it

may be wisest to plan the bilateral burr holes as a separate procedure since the existence of a membrane is by no means certain under these circumstances. In any case, burr holes should always be made eventually on both sides, even though the amount of fluid obtained on one side may have been negligible as compared with the other.

Craniotomy: The ultimate aim of surgical treatment is wide excision of both outer and inner subdural membranes if present. Craniotomy is routinely performed under endotracheal ether anesthesia with a plastic catheter placed in an ankle vein for constant infusion of fluids and blood. These operations should be carried out expeditiously with a minimum of operative trauma and blood loss and with adequate precautions against loss of body heat (p. 432).

A medium sized fronto-temporo-parietal osteoplastic flap is elevated in routine fashion after a preliminary burr hole has disclosed the existence of a membrane (Figure 176). This flap should extend forward to the hair line and medially to within one inch of the mid-line. In young infants, the bone is usually cut between the burr holes with stout scissors. The dura is opened in a circular fashion at the limits of the bony exposure. The outer membrane may be closely adherent to the dura; it can be separated and excised either as the dura is elevated or after the dural flap and membrane have been raised together (Figure 177). All solid clot and fluid contents of the hematoma are removed by suction and irrigation (Figure 178). The inner membrane varies a good deal in thickness and distribution. It is usually closely applied to the cortex and may occasionally be difficult to separate. This inner membrane, however, should also be widely removed. It is usually possible to excise both outer and inner membranes well beyond the limits of the exposure by careful dissection underneath the dural margins. In the average patient it is possible to remove membranes from within a centimeter of the falx down to the floor of the frontal and middle fossae and from the frontal pole back to the posterior parietal region.

The dura is closed tightly and the bone flap and scalp are reapproximated in routine fashion. Drainage is not used. A small transfusion of whole blood is routinely administered during the latter part of the craniotomy. Even very small infants tolerate these operations well if they have been prepared adequately pre-operatively and supported properly during the procedure. There are no particular problems during the post-operative period. Head dressings should never be tight because of the relative ease of scalp necrosis in this age group. If recovery is satisfactory, craniotomy may usually be performed on the second side in six to 10 days.

Post-operative subdural taps are occasionally necessary. This is particularly true when large hematomas have been present for a long time causing considerable cortical atrophy so that a deep space remains after operation

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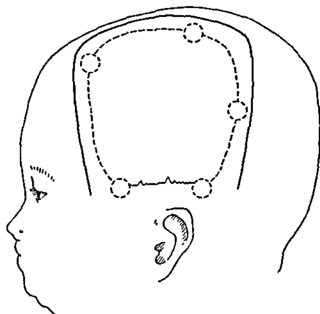


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Post-operative subdural taps are occasionally necessary. This is particularly true when large hematomas have been present for a long time causing considerable cortical atrophy so that a deep space remains after operation



Figure 177. Massive subdural hematoma in 6½ months old infant exposed at operation. Note the large collection of chocolate-colored fluid and fibrin seen through the translucent outer membrane.

into which the brain cannot immediately re-expand. If hemostasis has been complete at operation the fluid which collects in this space is only slightly xanthochromic and is not under increased pressure. When this is true, the fluid is tapped at increasing intervals of two, four, seven, 14 days until the amount becomes negligible. This may be done as an office or out-patient department procedure. If further bleeding occurs into this fluid, new membranes may form. In patients with extreme cortical atrophy the subdural space is never completely obliterated. Collapse of this space by subperiosteal removal of the bone flaps has been performed on one occasion in this clinic. When the brain expands up to the dural level at the time of operative removal of the membranes, there is usually no problem in post-operative fluid collection.

Results

On the regimen of treatment outlined, both case and operative mortality from subdural hematoma have been virtually eliminated in this clinic. Over 300 infants have now been treated according to this plan. All of this

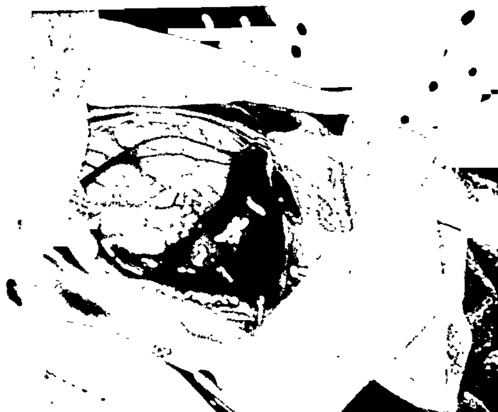


Figure 178. Typical recent subdural hematoma in a six months old infant. The thin outer membrane has been removed and a solid clot of blood partially organized is being reflected. There is no inner membrane at this stage. (Reprinted through the courtesy of The C. V. Mosby Company from *J. Pediat*, 24·1-37, 1944)

group have had subdural taps and burr holes performed. Three hundred and twenty-five craniotomies have been carried out in 222 patients under two years of age. Patients have now been followed up to 14 years.

The most significant factor in determining the end result has been the extent of cortical atrophy visualized at the time of operation. The retarded and deficient children have for the most part been those with large chronic hematomas with marked atrophy of the hemispheres already established by the time the membranes were removed. Of the entire group, between 70 to 80 per cent appear to be developing normally for their age or very close to it (Figure 179). Post-operative convulsions have been rare and residual neurological signs other than generalized hyperactivity of the deep tendon reflexes for a time have not been common. Re-formation of a subdural hematoma necessitating a second craniotomy for removal has occurred about 10 times.

Attention is called here to the peculiar problem of encapsulated collections of xanthochromic, high-protein fluid in the subdural space following meningitis, particularly due to *h. influenzae*, which is discussed elsewhere (p. 383).



Figure 179A (*Left*). Eleven year old girl who had bilateral subdural hematomas removed 10 years previously.

B (*Right*). Twelve year old boy who had bilateral subdural membranes excised during the first year of life.

Chapter VI

Injuries to the Spinal Cord

COMPOUND INJURIES involving the spinal cord are extremely rare in infancy and childhood. Closed trauma, however, occurs as a result of difficult delivery, vehicular accidents, and falls on the head and in the sitting position. These accidents may produce any degree of cord injury from simple concussion to complete crushing or anatomical transection. From a therapeutic point of view, the important injuries are those in which fracture or dislocation of the spine results in compression of the spinal cord or cauda equina and necessitates operative intervention or orthopedic manipulation for relief.

Concussion

Simple concussion occurs as a result of severe trauma in the neighborhood of the spine without actual compression or laceration of the cord itself. This is apt to occur when a child falls some distance landing flat on his back or is struck a sharp blow directly over the spine as in a kick by a horse. Such spinal cord concussion is accompanied by temporary and reversible interruption of function which does not require specific therapy as a rule.

Management includes only careful attention to the skin, bladder and bowels, and physical rehabilitation. Improvement often begins within a few hours and in children usually proceeds to complete recovery within a few days to weeks.

Birth Injury (Stretch Injury)

Since the ligaments and other soft tissues supporting the spinal column are very elastic at the time of birth, the cord may be severely stretched without any demonstrable skeletal injury. This type of diffuse stretch injury to the cord usually occurs in the low cervical or upper thoracic region.[25, 37, 38] There may be an actual disruption of the cord or more commonly diffuse intramedullary hemorrhage. It occurs almost invariably as a result of difficulties encountered in delivery of the after-coming shoulders and head in a breech extraction. Treatment, as with concussion, is limited to supportive measures. When physiological cord transection appears complete right after birth in the absence of skeletal deformity or fracture, the prognosis for recovery of function is extremely grave.

Combined Skeletal and Cord Injury

Three types of injury to the vertebral column are apt to damage the spinal cord:

Compression fracture of a vertebral body with acute angulation of the cord at the level of the spinal lesion.

Fracture-dislocation, commonest in the cervical region, and particularly prone to produce either transient or persistent cord compression.

Depressed fracture, producing cord injury when comminuted fragments of laminae, pedicles or body are driven into the spinal canal (Figure 180).

First-Aid and Transportation

It is important to recognize spinal injury as quickly as possible after an accident in order to avoid the possibility of increasing the degree of spinal cord damage by injudicious movement or manipulation. Spinal injury may be indicated by pain, postural deformity, tenderness, or by neurological changes in the trunk and extremities. If spinal injury is suspected, the child should not be allowed to sit, stand or walk. He should be lifted or rolled onto a stretcher or solid support for transportation in such a manner that the spine is not flexed, extended or twisted. It is wise for one attendant to maintain gentle traction on the child's head during all lifting and turning, particularly if he is frightened and crying.

If there is a cervical spine injury, the child should be transported on his

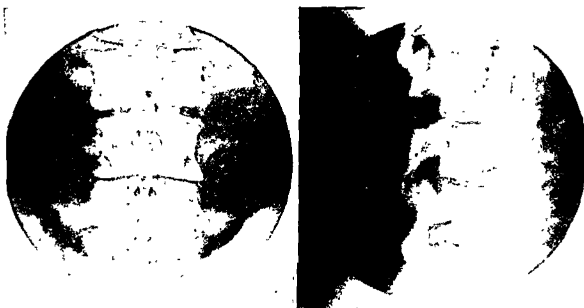


Figure 180 Antero-posterior and lateral roentgenograms of the lumbar spine of a 16 year old boy showing extensive comminuted fracture of the body, pedicles and laminae of L_1 with posterior displacement of bone into the spinal canal. Patient had a paraplegia below this level. Complete recovery followed slowly after open reduction and spinal fusion.

back with a folded coat or blanket under the shoulders to allow the head to fall back in slight extension. The sides of the head should be supported to prevent turning. If there is a thoracic or lumbar spine injury, the child may be transported in the prone position with a pillow under the head, or on his back with a support under the spine at the site of injury.

Examination

Since, both in infants and in uncooperative, frightened children, there may be no possibility of obtaining an adequate history, careful objective examination becomes all-important. Visible deformity of the spine should be noted together with local edema, ecchymosis, or contusion. The site of injury can often be determined by sliding a hand gently under the patient and examining along the spine for tenderness or palpable deformity. Distension of the bladder should be determined.

Neurological examination of the trunk and extremities of a recently injured child is fraught with many vicissitudes. However, with patience and perseverance, a satisfactory motor, sensory and reflex appraisal can usually be made. Below the level of cord transection there is flaccid paralysis, areflexia, anesthesia, and absence of vasomotor responses immediately after injury. If transection has occurred, the level is determined most accurately by moving from caudal toward cephalic dermatomes with a painful stimulus such as pin-prick.



Figure 181. Characteristic position of infant following high thoracic cord injury at birth. Note frog-like position of the legs and underdeveloped thoracic musculature with flaring of the rib margins. Intercostal respirations were completely paralyzed.

The paraplegic infant as a result of cord transection at birth lies in a characteristic position with the legs limp and everted in a frog-like position (Figure 181). There is no involuntary motor response or cry of pain in response to stimulus. The bladder soon becomes palpable.

X-ray examination of the entire spine should be carried out as soon as a good air-way is assured, the child is out of shock, a preliminary neurological

examination has been made, and sufficient personnel are available to move and position the patient carefully on the x-ray table. X-ray films are examined for contour and alignment of the vertebral bodies, linear or comminuted fractures of the laminae, pedicles or neural arches, and displacement of bone fragments into the spinal canal. It should be remembered that particularly in young children dislocation may be reduced spontaneously with residual cord damage but no deformity visible on x-ray examination. Also, it should be remembered that in the stretching type of cord injury occurring with breech delivery, there is usually no abnormality noted on x-ray examination.

Treatment

The surgeon must always keep in mind the danger of producing further injury to the spinal cord by improper treatment of a skeletal injury.

(1) Simple fracture of a vertebra without compression or displacement, with or without neurological involvement, is treated by immobilization in neutral position in a plaster body spica for at least six to 10 weeks.

(2) Compression fracture of a vertebral body is treated by hyperextension of the spine at the level of injury with fixation in this position until healing occurs. These fractures are not common in infants and children. General anesthesia is usually necessary to achieve proper hyperextension and apply a satisfactory body cast. When there is cord injury with any motor and sensory impairment, the plaster cast should be bi-valved at once to permit skin care and physiotherapy.

(3) Fracture-dislocation in the cervical region is associated with a high mortality when resulting from birth injury, particularly if there is sufficient cord damage to produce an immediate quadriplegia and paralysis of intercostal respiration. Skeletal or halter traction of the head in this age group is exceedingly difficult to maintain. In older children with low cervical fracture-dislocation, as from diving accidents, standard Crutchfield tong type of skeletal traction can be instituted. In infants and younger children where the skull is too thin for these tongs, wires or aluminum bands may be passed from one burr hole to another between the dura and the skull (Figure 182) Skeletal traction for cervical fracture-dislocation should probably never be discontinued in less than eight to 10 weeks, and then the patient should always have further support in a brace or cast for three to six months.

When cord compression is evident, if skeletal traction cannot be satisfactorily maintained or if it does not result in reduction of the fracture-dislocation within a few days, operative reduction should be attempted, followed by spinal fusion.

(4) Depression of comminuted bone fragments into the spinal canal at



Figure 182. Fracture dislocation of the cervical spine due to birth trauma. Reduction of dislocation accomplished by cervical traction. Tantalum band passed between two burr holes in each parietal region is attached to a yoke for pulley traction. The head of the bed is elevated to provide counter-traction.

any age should be treated by early laminectomy. Exposure must be carried out with caution in the area of fracture so as not to produce further cord damage. All loose bone fragments, displaced soft tissues and blood clot should be removed from the spinal canal. The dura is not opened unless it has already been lacerated. Tears in the dura caused by penetrating bone should be tightly closed. If there is destruction of articular facets, primary spinal fusion should be considered.

Skin Care

The paralyzed child should be placed as soon as possible on a foam rubber or air mattress with great care to protect the sacrum, trochanters, heels and other pressure points. The position of the paralyzed parts should be altered at least every two hours. Whenever available, a Stryker type frame should be used (Figure 183). All of the anesthetic skin should be washed, dried and lightly massaged daily. Bed coverings should be kept dry and smooth. Frequent tincture of benzoin application helps protect the skin in areas of pressure. If decubitus lesions develop, they must be kept scrupulously clean and free from any friction to promote healing.

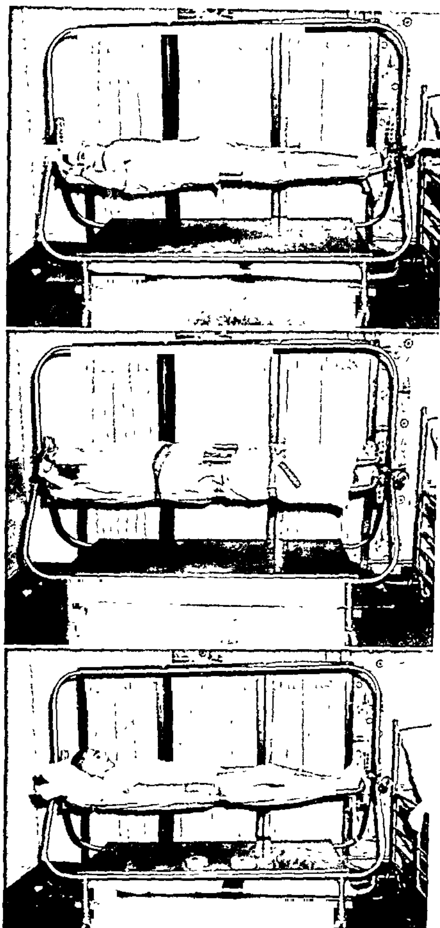


Figure 183 (*See legend on facing page.*)

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Excision of decubitus ulcers with plastic closure or skin grafting may occasionally be indicated.

Care of Bladder and Bowel

Since retention of urine develops at once after severe cord injury at any level, it is important to insert an in-lying urethral catheter under aseptic precautions within a few hours after injury. In small children, care must be exercised not to use too large a catheter. Repeated catheterization should be avoided. If prompt recovery of function does not occur, a plan of tidal drainage which provides automatic periodic drainage and irrigation of the bladder should be established. The height of the siphon curve in this system is determined by periodic cystometry as recovery proceeds through the stages of autonomy and automaticity.

Since retention of feces also occurs with severe injury to the cord, enemas should be started early and continued at regular intervals to promote automatic function. Fecal impactions develop easily. In cord injuries of less severity, abdominal distention due to ileus is a common complication in the first few days after injury. In the average child this can be relieved with small doses of prostigmine, enemas, and in-lying rectal tubes; Miller-Abbot tube decompression may be necessary occasionally.

Rehabilitation

The later stages of management of the paraplegic child are not within the scope of this book. However, it should be stated that physical rehabilitation should begin as soon as possible after injury. This includes proper positioning of the extremities to prevent deformity and early, regular, active and passive physiotherapy. Nutrition should be maintained at an optimum level at all times. Personal reassurance and encouragement by the doctor and by the child's family and friends are all-important.

Herniated Intervertebral Disc

Acute rupture of the annulus fibrosis with protrusion of the nucleus pulposis of an intervertebral disc in the lumbar region has been reported

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Figure 183. Stryker frame in use for treatment of 16 year old boy with comminuted fracture dislocation of the spine at L_2, with paraplegia below this level Open reduction and spinal fusion have been performed.

A (*Upper*). Patient in the prone position in anterior plaster shell.

B (*Middle*). Patient ready for turning with both plaster shells in position and both halves of the Stryker frame secured with straps.

C (*Lower*) Patient turned to supine position. Anterior shell has been removed. Posterior plaster shells also maintain the feet in right angle dorsi-flexion.

in children rarely. In the presence of low-back pain accompanied by unilateral leg pain of sciatic distribution and by neurological evidence of nerve root compression in childhood, the diagnosis of herniated intervertebral disc should be considered. In this clinic, the youngest patient with this lesion was a girl who had just passed her thirteenth birthday. She exhibited paravertebral muscle spasm, sciatic nerve tenderness, limited straight leg raising, limited motions of the lumbar spine, weakness of plantar flexion of the foot, absent ankle jerk and hypalgesia over the S_1 dermatome.

Myelogram revealed a defect at the lumbo-sacral interspace (Figure 184), and interlaminar removal of a large herniated disc was followed by complete relief of symptoms and early resumption of normal activity.

Herniated intervertebral disc protrusion in the cervical region has not been seen in children in this clinic.



Figure 184 Antero-posterior myelogram showing unilateral defect at the lumbo-sacral interspace due to a massive herniated intervertebral disc in a girl of 13 6/12 years

Chapter VII

Injuries to the Peripheral Nervous System

BRACHIAL PLEXUS INJURY

Clinical Features

Two types of injury to the brachial plexus are seen in infancy and childhood. By far the commonest is that occurring at birth due to stretching of the plexus as a result of strong traction on one upper extremity during delivery of the after-coming head in a breech presentation. The other type of injury usually occurs in falls, as from a moving vehicle, where one arm is caught or grasped and this shoulder forcibly drawn away from the head and neck.

Birth injury to the brachial plexus is often associated with other evidence of trauma, particularly fracture of the clavicle or surgical neck of the humerus. It is almost always unilateral and is usually recognized at once since the paralysis is most severe immediately after delivery.

The appearance of the involved extremity depends upon which part of the plexus is injured. The commonest type of paralysis involves the upper arm, so-called Erb's palsy, due to injury of the fifth and sixth cervical roots, or upper trunk of the plexus. In this condition the infant lies with the shoulder in adduction and internal rotation, the elbow in extension, the forearm in pronation and the wrist in flexion (Figure 185). The paralyzed muscles include the deltoid, spinati, biceps, brachialis, brachioradialis, and to a less extent the extensors of the wrist and fingers. There may be paralysis of the ipsilateral leaf of the diaphragm. The biceps and radio-periosteal reflexes are absent. Sensory changes are difficult to elicit.

Much less commonly, paralysis involves principally the lower arm and hand, so-called Klumpke's paralysis, due to injury to the seventh and eighth cervical roots or lower trunk of the plexus. Here the weakness involves the flexors of the wrist and fingers and the intrinsic muscles of the hand so that the latter is flat, edematous and without ability to grasp (Figure 186). There may be an ipsilateral Horner's syndrome due to injury to the first thoracic root as well. Sensory loss is demonstrable, particularly on the volar surface of the hand.

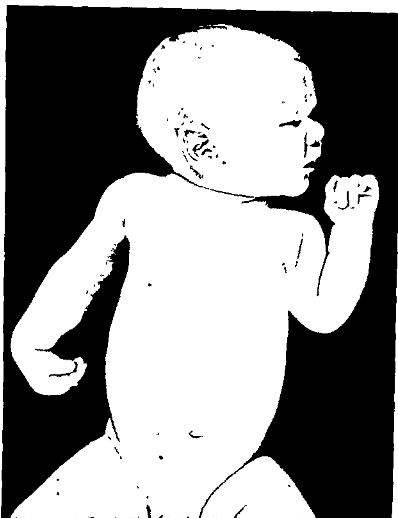


Figure 185 New-born infant with right brachial plexus palsy of the Erb's type due to birth injury. Note extension of the elbow, adduction and internal rotation of the shoulder and flexion of the wrist.

In extremely severe injuries, most of the plexus may be involved resulting in paralysis of the entire arm (Figure 187). The arm hangs limply from the shoulder, is areflexic and anesthetic up to the middle of the upper arm.

Brachial plexus injury may consist of actual avulsion of the anterior roots from the spinal cord, of severe stretching of the roots or trunks of the plexus, or of hemorrhage into or around the plexus. When the roots are avulsed from the cord there is usually some bleeding into the subarachnoid space. Consequently, blood in the spinal fluid on lumbar puncture shortly after birth in patients with brachial palsy carries an ominous prognosis for return of function. Also if a Horner's syndrome is present, it usually indicates an intraspinal avulsion of roots. In the commoner type of upper arm paralysis, the maximum injury occurs at the point where the fifth and sixth nerves join to form the upper trunk of the plexus. Here there may be con-

siderable hemorrhage into and about the nerves. Examination of the patient may show ecchymosis and soft tissue swelling in the supraclavicular fossa in the acute phase and diffuse induration later on.

Treatment

Surgical treatment of birth injuries to the brachial plexus is discouraging. Fortunately by far the greater number of these paralyses recover spontaneously. This is true also of the milder stretch injuries due to falls. During the period of observation awaiting spontaneous return, it is important to prevent deformity due to overaction of muscles whose antagonists have been paralyzed and to carry out regular passive physiotherapy to prevent atrophy of the involved muscles. In the common upper arm type of palsy, the arm should be kept abducted and externally rotated at the shoulder, flexed to ninety degrees at the elbow and extended at the wrist. This is accomplished by a so-called "airplane" splint, which is well tolerated by even small infants if carefully fitted (Figure 188).



Figure 186. Eighteen months old boy with lower arm type brachial palsy and co-existent Horner syndrome. Note the narrow palpebral fissure on the right and normal position of the upper arm.

In this clinic exploration of the brachial plexus has been carried out on patients with injuries due to falls or other forms of post-natal trauma and on those birth injuries showing no progressive improvement after three to six months in whom there was palpable thickening in the supraclavicular fossa or in whom there was definite history of swelling or ecchymosis over the plexus in the acute phase of injury. Many of these injuries show progressive spontaneous improvement for a number of months and surgical exploration should probably be delayed as long as this is true.

The most that one can hope to accomplish at operation usually is careful neurolysis, so that unless there is extrinsic scarring subject to removal by meticulous dissection, it is better not to risk the danger of additional scar-

ring by extensive exploration. Avulsion of the roots intradurally or in the intervertebral foramina does not lend itself to surgical repair.

Exploration of the brachial plexus in infants is carried out through a transverse incision in the line of the natural neck crease about half way between the clavicle and the angle of the jaw (Figure 189). The roots are identified as they emerge between the anterior and middle scalene muscles and join to make up the trunks of the plexus. A stimulator capable of delivering graduated faradic current should be available throughout the operation. If there is avulsion of the trunks or cords of the plexus and additional exposure is needed to perform an anastomosis, it should be obtained by extending the lateral end of the original incision downward over the apex of the shoulder, dividing the clavicle and attachents of the pectoral muscles and freeing up the nerve trunks distally.

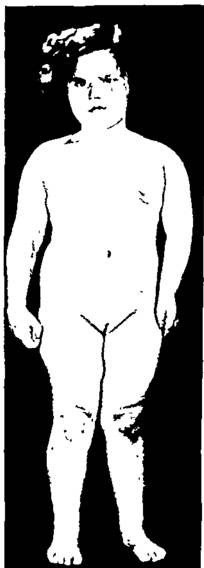


Figure 187. Six year old girl with left whole arm type of brachial plexus paralysis due to birth injury. Note limp position of left upper extremity which is smaller than the right. There is a Horner's syndrome on the left.

The results of neurolysis in birth injuries are difficult to assess and are discouraging. However, there have been instances where improvement has apparently followed operation to a degree greater than could reasonably be expected to have occurred spontaneously. The prognosis in whole arm and lower arm types of injury is much worse than in the upper arm variety.

Rehabilitation of neglected or unimproved brachial plexus injuries by various orthopedic procedures is beyond the scope of this discussion but should be pursued at the proper time in every patient in whom no further recovery of neurological function is anticipated.

FACIAL PARALYSIS

Partial or total paralysis of the facial nerve is occasionally a complication of delivery. This paralysis is of the peripheral or infranuclear type and usually results from undue pressure of a forceps blade over the nerve somewhere between its emergence from the skull at the stylo-mastoid foramen and its points of entry into the facial muscles. It is usually unilateral and usually involves all of the musculature of



Figure 188. Two week old infant with upper arm type of brachial palsy on the right. Patient has "airplane" splint in place to maintain proper position during observation for spontaneous recovery.

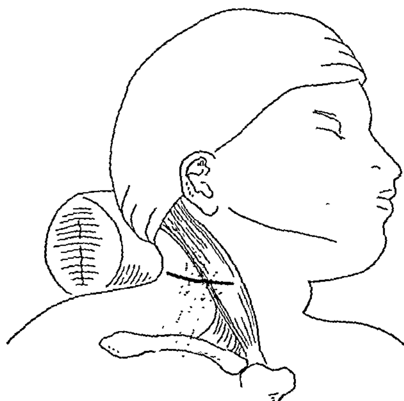


Figure 189. Location of incision for exploration of the brachial plexus in infants and children. Incision is half-way between clavicle and angle of the mandible. Mid-point of the incision lies over the lateral border of the sternomastoid muscle.

one side of the face, though not necessarily to an equal degree. Other evidence of difficult delivery is often presnt, such as cephalhematoma, depressed skull fracture, subarachnoid hemorrhage, or extreme moulding of the head.

The clinical appearance of this injury is characteristic (Figure 190). At rest the face may be symmetrical but when the child cries the lack of movement of the facial musculature on the affected side is readily apparent. The infant is unable to close the eye-lids on this side and the eye-ball turns upward. There is sagging of the corner of the mouth with drooling of oral secretions from this side. If the paralyzed side is placed uppermost, these infants suck well since the tongue is not paralyzed and they will not drool in this position.

These injuries are due ordinarily to compression and not laceration of the nerve. *Fortunately in most instances the damage to the nerve is* fairly mild and recovery begins early. If there is any evidence of return of function within two or three weeks, it usually proceeds steadily to complete recovery within two to three months. In more severe degrees of compression, return of function may not begin for several months, but even under these circumstances the prognosis for return of function eventually



Figure 190 Birth injury to the left facial nerve caused by obstetrical forceps.

A (*Left*). Note lack of motion of the entire left facial musculature when the baby cries.

B (*Right*). The point of injury to the facial nerve is seen in the pre-auricular region.

is excellent. Rarely, peripheral degeneration is apparently complete and permanent.

Surgical intervention is usually not indicated in the early phase of these injuries. Since this is a compression injury without local hemorrhage or laceraton, there is nothing to be gained by exploration of the nerve. Anastomosis to other cranial nerves should not be considered because most of the lesions recover spontaneously. In the rare patient who shows no recovery in several years, anastomosis might be considered but after this prolonged denervation such muscles cannot be expected to respond well to a new nerve supply. Various types of plastic procedures may be employed to support the paralyzed side of the face at rest if it is asymmetrical in these unimproved patients. If there is a laceration of the cheek and the nerve has actually been divided, it should of course be sutured directly (Figure 191).



Figure 191 Same patient as shown in Figure 190, two years and 10 months following suture of lacerated facial nerve.

The mother or nurse should be instructed in care of the eye in infants with facial paralysis. The eye should be irrigated two or three times daily at least and protected in the interim with bland ointment. Light massage with the tips of the fingers of all the paralyzed face muscles should be carried out regularly until full recovery takes place. Attempts at holding up the cheek and corner of the mouth by adhesive strapping in young infants are usually futile.

PERIPHERAL NERVE INJURIES

Aside from birth injuries to the facial nerve and to the brachial plexus, trauma to the peripheral nervous system in infants and children is not common. The types of injury occasionally seen are similar to those encountered in adults in civilian practice, such as division of the ulnar and median nerves about the wrist due to lacerated wounds, division of the

radial nerve in the lower third of the arm associated with fracture of the humerus, of the ulnar nerve in fractures of the elbow, and of the common peroneal nerve associated with fracture of the neck of the fibula. In this clinic the principles emphasized in World War II of early exploration and neurolysis, of delayed primary suture and of anastomosis without tension using only neurilemmal sutures of fine tantalum wire (.003 inch) have been followed (Figure 192). In children, the regeneration of axons once the suture line has been bridged is very rapid, so that recovery of function may be expected in much less time usually than following a similar injury in an adult. Post-operative splinting and physical rehabilitation are equally as important as in adult injuries.

Neurolysis After Peripheral Nerve Injury Due to Faulty Intramuscular Injections

One of the hazards of increased intramuscular administration of antibiotics has been the greater incidence of peripheral nerve injury due to faulty injection near or into one of the major nerve trunks. This unfortunate complication is most apt to occur in an uncooperative, squirming infant. The



Figure 192A (*Left*) Lateral roentgenogram of the elbow showing severe supracondylar fracture of the humerus in an eight year old boy associated with complete paralysis of the radial nerve.

B (*Right*). Lateral roentgenogram one year later showing solid healing of fracture in good position. Note position of tantalum wire sutures at the point of anastomosis of the severed radial nerve Patient had close to 100 per cent functional return in less than a year.

nerves most commonly involved are the radial due to deltoid muscle injection placed too low and the sciatic due to a gluteal injection placed in the medial or lower part of the buttock instead of the upper, outer quadrant. Interruption of function is probably due to a high local concentration of the agent which produces intense intraneural and perineurial reaction.[22]

Experience in this clinic[133] suggests that recovery of function lost due to local neuritis is facilitated by early extensive neurolysis of the resultant scarring (Figure 193). While it is always tempting to pursue conservative treatment longer in the hope of spontaneous recovery, there is little to be lost and possibly much to be gained by early careful exploration in the region of injury. At operation the nerve trunk should be completely mobilized from adhesions to surrounding tissues and all constricting scars in the nerve itself should be divided longitudinally in such a way as to preserve the integrity of the axons. It is felt that this type of early lysis of perineurial and intraneural scarring will be followed by functional return that is accelerated in rate and probably increased in final extent as well. It is recommended that if there is no clinical evidence of return of function within two to three weeks of onset of paralysis apparently due to a faulty injection, exploration of the involved nerve accompanied by thorough neurolysis be performed without delay.



Figure 193 Extent of sciatic nerve that required neurolysis because of reaction following faulty intramuscular injection of streptomycin. (Reprinted through the courtesy of The Massachusetts Medical Society from *New England J. Med.*, 242:973-975, 1950.)

PART IV

INTRACRANIAL TUMORS

Chapter I

General Considerations

THERE ARE several features of intracranial neoplasms in infancy and childhood worthy of general comment before proceeding to a more detailed discussion of the characteristics and treatment of each individual type of tumor. In the first place, intracranial neoplasms in childhood are not uncommon. It has long been recognized that there is a peak in the incidence of brain tumors toward the latter part of the first decade of life.[10, 45, 72] They occur, though comparatively rarely, in the first year of life, increase in frequency up to five to eight years of age, and then fall off as adolescence is approached. In the neurosurgical department of The Children's Medical Center, Boston, approximately 30 to 40 patients under the age of 12 are treated each year for intracranial tumors. This is a little less than 10 per cent of the children admitted to the hospital for neurosurgical treatment. The ratio of males to females is not significant; 160 males to 153 females.

Two of the more distinguishing features of intracranial tumors in childhood as distinct from those in adults relate to their histological type and to their anatomical location.

Tumor Type

It is of interest to note that in 1890, Starr [187] estimated from pathological studies that at least 50 per cent of all intracranial tumors in infancy and childhood were tuberculomas. As late as 1925, Critchley [36] reported from England that among children tuberculomas were the most common and gliomas second in frequency amongst brain tumors. It is a tribute to the medical and public health management of tuberculosis that these lesions are now a rarity in most parts of the world.

The large proportion of gliomas to other types of tumors is perhaps the most significant characteristic of the pediatric age group. In 1927, Cushing [42] discussed his experiences with the tumors of preadolescence and found that 75 per cent of 154 tumors were gliomas. Bailey, Buchanan and Bucy [10] also found that 75 per cent of tumors in 100 children under 15 were gliomas; Smith and Fincher [184] reported 86 per cent gliomas among 100 tumors; Keith, Craig and Kernohan [108] reported 84 per cent among 427 patients; Walker and Hopple [200] reported 78 per cent gliomas among 100 children; Cuneo and Rand [39] reported 70 per cent among 83 patients. In

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our own series, which is limited to children under 12 years of age, the proportion of gliomas among 313 patients is 80 per cent (Figure 194).

These remarkably consistent figures from different clinics compare with an average of only 45 per cent gliomas among large series of intracranial tumors in adult patients. This discrepancy is accounted for largely by the great frequency of the mid-line cerebellar gliomas in children which are uncommon in adults and by the almost complete absence in childhood of three of the common tumor types found in adult life; namely, the meningiomas, acoustic neurinomas and pituitary adenomas.

Anatomical Location

In adults approximately 70 to 75 per cent of intracranial neoplasms occur above the tentorium, whereas in childhood just the reverse is true. Thus, tumors in the cerebellum, pons, medulla, fourth ventricle, and cerebello-pontine angles account for 60 to 70 per cent of all the intracranial tumors in the pediatric age group. In our series of 313 tumors, 128, or 40 per cent, were supratentorial, and 185, or 60 per cent, infratentorial.

There is a tendency for tumors in early life to occur along the central neural axis; that is, within the third or fourth ventricles or within or attached to the brain stem. This location results in a lack of localizing neurological signs, but frequently in early obstruction to the normal circulation of spinal fluid. As a result, early symptoms and signs of these lesions are more apt to relate to increased intracranial pressure than to any abnormality of neurological function.

In infancy the ordinary clinical signs of increased intracranial pressure may be delayed in appearance because of the ability of the skull to expand. *Intracranial pressure at any given moment is a function of the relation of the size of the intracranial chamber to the amount of its contents. The* contents consist of brain, spinal fluid and blood. If the mass of the brain increases in amount by growth of a neoplasm, or the quantity of spinal fluid increases as a result of obstruction to its circulation or absorption, then intracranial pressure will increase unless the capacity of the cranium can enlarge proportionately. Before the cranial sutures have fused firmly such expansion of the cranium is possible; therefore, a tumor mass may achieve considerable proportions or the degree of hydrocephalus become severe before an infant exhibits clinical evidence of increased pressure. Under these circumstances, of course, enlargement of the head at greater than the normal growth rate may be the only clue to the pathological process.

The paucity of localizing neurological symptoms and signs, the delayed appearance of increased intracranial pressure due to expansion of the head, and the inability of infants and young children to define complaints accurately all serve to make early detection of brain tumors in this

age group difficult. As will be stressed in subsequent sections, this makes the wide employment of objective diagnostic procedures in childhood essential.

The various intracranial tumors of childhood will be discussed under the following headings: (1) posterior fossa tumors, including particularly cerebellar astrocytomas, medulloblastomas, ependymomas and gliomas of the pons; (2) gliomas of the cerebral hemispheres; (3) craniopharyngiomas; (4) intracranial dermoids and teratoid tumors; and (5) miscellane-

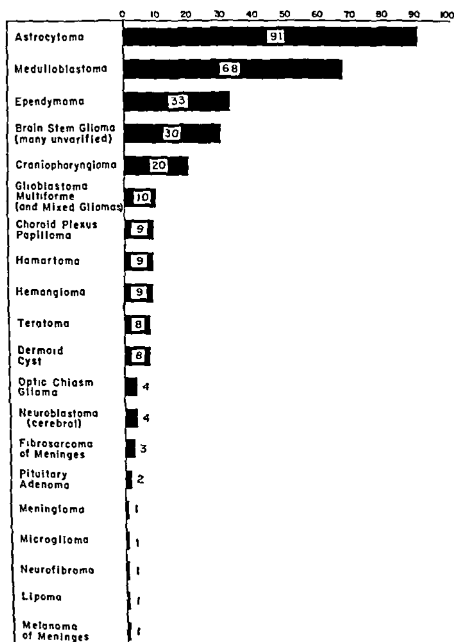


Figure 194. Frequency of tumor types found in 313 consecutive intracranial lesions in children under 12 years of age.

ous tumors, including choroid plexus adenomas and tumors of the skull and meninges. We have no explanation other than circumstance why in this particular series no patients under 12 years of age have been encountered with cystic hemangioblastoma of the cerebellum or with pinealoma.

Chapter II

Posterior Fossa Tumors

THE PREPONDERANCE of tumors below the tentorium in infancy and childhood has already been mentioned. Sixty to 70 per cent of all intracranial new growths in this age group occur in the posterior fossa. Because this is a relatively small space, a tumor anywhere within the posterior fossa may soon involve any of the vital structures present. Whereas localization of function within the cerebrum is well developed and often permits accurate clinical localization of a neoplasm, this is not true in the cerebellum. Also, because of the narrow pathways through which spinal fluid must pass within the posterior fossa, namely the aqueduct, fourth ventricle and basilar cisternae around the medulla, a tumor anywhere below the tentorium may very early cause obstruction to spinal fluid circulation with resultant internal hydrocephalus. Thus, the actual clinical picture produced by a tumor in the posterior fossa, whether medulloblastoma, astrocytoma, ependymoma, hemangioblastoma, dermoid cyst, or choroid plexus adenoma of the fourth ventricle may be quite uniform. This clinical picture will be discussed in a general way before proceeding to individual types of tumor.

Symptoms and Signs

The symptoms and signs of posterior fossa neoplasms as a group are due primarily to increased intracranial pressure and secondarily to local compression of the cerebellar nuclei and the brain stem. There is nothing in the clinical history or physical examination which makes the histological diagnosis conclusive. It is true that in medulloblastomas the history tends to be shorter and it appears to progress relentlessly, while in astrocytomas the course tends to be longer with intervals of comparative symptomatic freedom. However, these criteria are not reliable, and in any given patient at any given moment the cell type cannot be predicted accurately previous to operation.

Twenty years ago Cushing[44] devised a composite history of children with cerebellar astrocytomas which is still appropriate to a vast majority of all posterior fossa neoplasms and is repeated here.

"A child apparently normal in all respects begins toward the end of the first decade, possibly after a fall or an attack of whooping cough, to have

early morning headaches with vomiting. Nothing much is made of this by the family doctor, should he be called in, for the child subsequently feels perfectly well, has had breakfast and wants to go out and play. This daily performance may continue for a considerable time, the child even going to school meanwhile. There may then be a remission of weeks or perhaps months and the episode be forgotten. On their re-occurrence, the symptoms are likely to be more pronounced and are apt to be ascribed to some gastro-intestinal disturbance. This appears the more probable since the child finds that straining at stool brings on a headache and there is a tendency to become constipated. What is more, a mild daily laxative usually serves completely to mask the symptoms.

This sort of thing continues off and on until it becomes evident that the child is a little clumsy at play and gets knocked over easily. Very possibly, ere this, the periodic headache and vomiting will have ceased completely or at least have occurred at much longer intervals; and if parents are observant they may notice that the child's head in the interim has increased in size more rapidly than it should. This, however, is usually discounted for the child meanwhile has become free from complaints and in all respects appears alert and well.

Matters may run on in this way for an indefinite time, possibly with some increase in clumsiness of movement or in some instances with no noticeable change whatever until it suddenly becomes apparent, perhaps at school, that the child's sight is poor. To counteract this glasses are usually prescribed; but even should an ophthalmoscope be resorted to, a child's retina is less easily examined than that of an adult and, because of the decompressive effects of the enlarging head, the optic papillae often show no measurable swelling and the fact of their being pale and with margins blurred may easily pass unrecognized."

The important symptoms and signs of posterior fossa tumor include: headache, vomiting, unsteadiness of gait, weakness, strabismus, nystagmus, seizures, papilledema, enlargement of the head, and stiffness of the neck.

Headache: This usually begins insidiously. In children who are old enough to complain accurately it may occasionally be limited to the occipital region, but is usually generalized. In infants headache from increased intracranial pressure may be manifested only by restless irritability and persistent desire not to be handled. The headache is often intermittent, particularly early in the course of the disease and often seems to be most severe in the early morning.

Vomiting: Perhaps the commonest symptom associated with posterior fossa tumors, and certainly one present in children of all ages, is vomiting. This is presumably a manifestation of increased intracranial pressure for the most part but may in some measure also be due to direct compression of vagal nuclei in the medulla oblongata. This vomiting is often forceful,

but is only rarely projectile, particularly early in the progress of the lesion. Vomiting is commonly misinterpreted as a symptom of some other ailment because of its frequency in many varieties of childhood illness. For some reason which is not clear vomiting is particularly common in the morning, at the time of or following the first meal of the day. The patient may often be able to eat again immediately. As the disease progresses, vomiting of practically everything taken by mouth may occur causing severe malnutrition.

Gait Disturbance: Usually the first manifestation of ataxia noted is a tendency for the child to fall frequently, to run into furniture, or to appear as "though he were drunk" on ordinary walking. When examined subsequently the child is noted to walk with a lurching gait on a wide base (Figure 195). He is unsteady in the Romberg position and eventually may be unable to stand at all, even with support. This is primarily a truncal ataxia; that is, there may be marked unsteadiness in standing and walking while at the same time if the child is lying down, he will show little or no ataxia or dysmetria of the extremities.

Weakness: Easy fatiguability often becomes apparent as an early symptom. The child may want to continue playing but simply have to stop because of generalized weakness. This often progresses to extreme hypotonia, particularly of the lower extremities. Associated with this there is marked diminution to absence of the deep tendon reflexes, again particularly in the lower extremities. Malnutrition from repeated vomiting may, of course, contribute to the generalized motor weakness (Figure 196).

Strabismus: As a result of internal hydrocephalus, paralysis of one or more of the cranial nerves supplying the extra-ocular muscles often occurs. Most commonly this is a unilateral sixth nerve weakness. The paralysis of the external rectus which this produces is manifested by an internal squint, and if the patient is old enough to note it, by diplopia (Figure 195). Third nerve palsy as a result of increased pressure occurs less often.

Nystagmus: Although a classical sign of cerebellar disease, nystagmus is often absent until late in the course of posterior fossa tumors in children. There may be marked truncal ataxia with no nystagmus. Certainly, absence of this sign never justifies assumption that a cerebellar tumor can be ruled out. When present, nystagmus is usually lateral and accentuated on forced lateral gaze. Vertical nystagmus occasionally occurs with lesions of the anterior cerebellum and aqueductal areas.

Seizures: Convulsive seizures are uncommon in infratentorial tumors. Occasionally in advanced lesions, "cerebellar" seizures occur. These consist of sudden onset of stiffening of all the extremities into a decerebrate type of pattern and usually are accompanied by abnormalities of respiration and heart rate. The patient may become completely unresponsive, cyanotic and

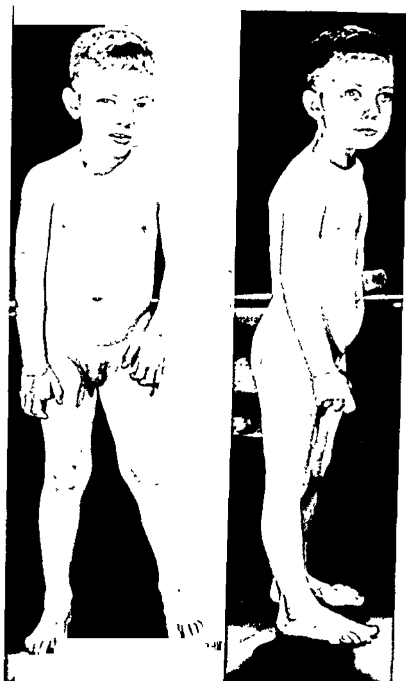


Figure 195. Four and seven-twelfths year old boy with midline solid astrocytoma of the cerebellum Note: (1) characteristic stance with feet placed widely apart, (2) rigidity of the neck and tilting of the head slightly toward the left, and (3) squint caused by sixth nerve paralysis.

rigid, with dilatation of the pupils and inability to swallow. Respiratory failure and death may occur in such an attack.

Papilledema: Since increased intracranial pressure is so prominent in posterior fossa tumors, it is to be expected that papilledema should be a common physical sign. In our experience the eye-grounds have been normal with extreme rarity among children with infratentorial tumors.



Figure 196 (*Left*). Two and three-twelfths year old boy who had been vomiting profusely for six weeks and had become too weak to stand. Patient had an ependymona of the fourth ventricle and improved rapidly after subtotal excision.

Figure 197 (*Right*). Four and five-twelfths year old girl with right cerebellar hemisphere astrocytoma. Note tilt of the head toward the right side. Patient refused to move her neck voluntarily out of this position.

This is true even in infancy where expansion of the head has permitted decompression. Unfortunately papilledema has often been extreme at the time of hospitalization, with secondary optic atrophy already present. In childhood, irreversible loss of vision develops with unusual rapidity in the presence of papilledema. It seems superfluous to reiterate here that careful ophthalmoscopic examination, in spite of its difficulty sometimes in small children, should be an integral part of every diagnostic investigation.

Enlargement of the Head: In infants with posterior fossa tumors, increase in size of the head may be the first abnormality noted. There is nothing characteristic about this enlargement to differentiate it from that

seen in hydrocephalus due to other causes (Figure 109). Usually the separated sutures may be palpated and on percussion a "cracked-pot" percussion note is elicited.

Stiffness of the Neck: When infratentorial tumors encroach upon the dura, fill the cisterna magna, or extend through the foramen magnum into the upper cervical canal, stiffness of the neck is a common symptom. The child may hold his head rigidly in one position to minimize pain and will resist efforts of the examiner to move his neck (Figure 197). If increased pressure has been present for some time, the occipital bone is often markedly thinned and there is unilateral or bilateral suboccipital tenderness.

Other late signs of increased intracranial pressure such as drowsiness, bradycardia, slowed respirations and hypertension need little comment here.

X-ray Examination

Routine x-ray films of the skull should include the following views: antero-posterior, postero-anterior, basilar and stereoscopic lateral. In the presence of infratentorial neoplasms during childhood these films will rarely be entirely normal.

In this age group the commonest abnormality noted is separation of



Figure 198. Antero-posterior and lateral roentgenograms of the skull showing the characteristic picture of severe acute increased intracranial pressure due to a tumor obstructing the fourth ventricle. Note marked separation of all the cranial sutures without any thinning of the posterior clinoid processes or unusual increase in the convolutional markings.

the cranial sutures, most marked usually of the coronal and then the sagittal (Figure 198). In infants, a soft tissue swelling in the region of the anterior fontanelle may be visible. When internal hydrocephalus becomes marked there is often erosion of the posterior clinoid processes due to compression by the dilated third ventricle (Figure 199). Asymmetry or thinning of the occipital bones is occasionally visible. With long-standing increased pressure there may be greatly accentuated convolutional markings on the inner table of the entire calvarium (Figure 199). The occipital bone should be examined carefully in the basilar view for evidence of mid-line defects that might indicate the path of a congenital sinus leading to a dermoid cyst (Figure 64).

Calcification is not common in posterior fossa neoplasms but is occasionally seen, usually in the basilar view. It appears as amorphous flecks and is most common in ependymomas or mixed gliomas (Figures 199 and 222).

There is no place for encephalography in the diagnosis of posterior



Figure 199. Lateral roentgenogram of the skull of a seven and a half year old boy with chronic increased intracranial pressure Note: (1) separation of the cranial sutures, (2) thinning of the posterior clinoid processes, (3) increase in convolutional markings, and (4) calcification within an astrocytoma of the cerebellum (arrow).

fossa neoplasms except possibly for visualization of the aqueduct and fourth ventricle in patients with suspected pontine gliomas who have no evidence of increased intracranial pressure (p. 266).

Ventriculograms are readily performed in most children with posterior fossa tumors by introducing a needle into a lateral ventricle through the separated coronal suture; when this is not possible, a drill or burr hole is made (Figure 200). However, ventriculograms are not made routinely. Ventriculography may be a very disturbing procedure in young patients with severe intracranial pressure. When performed, it should be done if possible under local anesthesia and preparations should always be made to follow with posterior fossa operation at once. If inhalation anesthesia is to be employed for craniotomy, it is often wise to reinsert a needle into the ventricle after the x-ray films have been made to relieve any pressure that may be present before induction of anesthesia is commenced.

Ventriculography is ordinarily limited to patients in whom the clinical diagnosis of posterior fossa neoplasm is open to doubt. If there is markedly increased intracranial pressure in a child with a staggering gait, diminished reflexes, hypotonia and no lateralizing signs, ventriculography is often omitted and ventricular estimation performed instead at the time of posterior fossa exploration. This procedure consists in introducing a needle into each lateral ventricle. If the ventricles are symmetrically dilated and under equally increased pressure, with free communication between them, it is assumed that the obstruction is in the fourth ventricle, aqueduct or posterior third ventricle, in all of which circumstances the proper procedure is exploration of the posterior fossa. If there is any asymmetry of the lateral ventricles, if the fluid obtained in one differs in any way from that of the other, or there seems to be any interventricular obstruction, then ventriculography is performed at this time.

Ventriculography in most posterior fossa neoplasms demonstrates symmetrical dilatation of the lateral and third ventricles and also the aqueduct

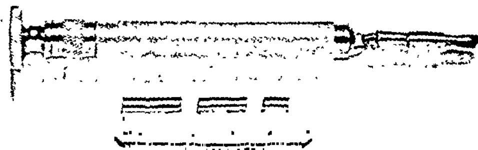


Figure 200 Simple hand drill with guard for penetration of the skull in children to perform ventriculography or therapeutic ventricular puncture. Such a drill is kept sterile in the operating room and on the ward at all times

if this is visualized. Occasionally air gets into the fourth ventricle and shows it shifted laterally (Figure 201), or the air may actually outline tumor projecting into the ventricle. Visualization of the aqueduct and fourth ventricle is facilitated by taking lateral films with the child in upside down position (Figure 202). Often the dilated aqueduct is seen telescoped on itself or displaced upward or forward (Figure 203). In large cerebellar tumors, usually cystic astrocytomas, elevation of the tentorium with displacement of one or both occipital horns of the lateral ventricles may be seen. It is impossible, however, *to make a reliable diagnosis of the tumor* type on the basis of ventriculography.



Figure 201. Postero-anterior ventriculogram of five year old child with large cystic astrocytoma of left cerebellar hemisphere. Note deformity and marked shift toward the right of the fourth ventricle.



Figure 202. Lateral upside down ventriculogram showing dilated lateral and third ventricles, a dilated distorted aqueduct, and invasion of the deformed fourth ventricle by cerebellar medulloblastoma (arrows).

Electroencephalography

The electroencephalogram is helpful in investigation of posterior fossa tumors occasionally in differential diagnosis from cerebral hemisphere lesions. It usually records the presence of increased intracranial pressure as evidenced by generalized slowing and increased amplitude. These changes are usually most marked in the occipital leads (Figure 204).

Treatment

Certain general considerations in the surgical treatment of posterior fossa tumors will be discussed here; specific problems relating to each type of tumor will be dealt with separately in subsequent sections. It is our feeling that every suspected posterior fossa neoplasm should be subjected to surgical exposure. No pre-operative criteria of tumor types are reliable for every case. Since the results of treatment are so good in astrocytomas of the cerebellum as compared with other infratentorial gliomas, satisfactory biopsy at least should always be performed to rule out one of these surgically favorable tumors. It is a terrible tragedy to submit a child with evidence of a mid-line cerebellar tumor to x-ray therapy with an assumed

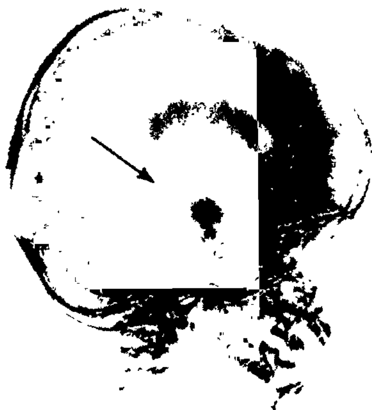


Figure 203. Lateral ventriculogram of a two year old child showing marked symmetrical dilatation of the lateral and third ventricles. Note obstruction in the upper portion of the aqueduct with failure of air to pass into the enlarged posterior fossa. Patient had an ependymoma of the fourth ventricle.

diagnosis of medulloblastoma, and then discover at operation later when pressure signs have increased and the patient has become blind that the lesion is in reality a cystic astrocytoma.[18, 94]

Exploration of the posterior fossa is performed in infants and children under endotracheal ether anesthesia in the face-down position using a horseshoe shaped rest to support the head (Figure 205). We have not employed the upright position for infratentorial operations in small children. With an endotracheal tube in place the head can be well flexed. A small roll under each shoulder and under the hips elevates the chest and abdomen sufficiently to permit an adequate respiratory excursion.

In infants and young children we routinely employ a mid-line skin incision (Figure 206). The occipital muscles are separated subperiosteally from the occipital bone and the arch of the atlas. If a transverse incision is made in these muscles for 2 to 3 cms. from the mid-line about a centimeter below their occipital insertion adequate exposure of the entire

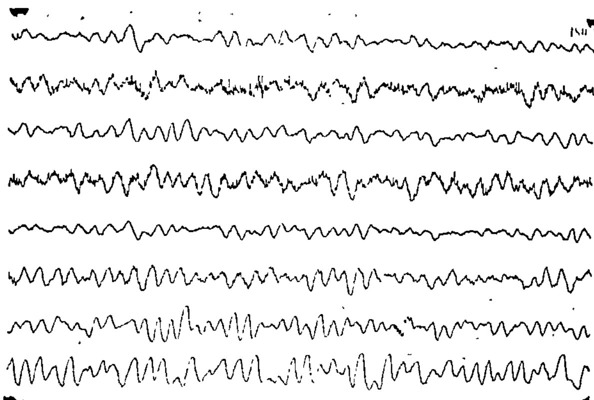


Figure 204 Electroencephalogram in one year old infant with cystic astrocytoma of the cerebellum. Note characteristic bilateral slowing and increase in amplitude in both occipital leads (lower two lines).

posterior fossa is usually possible. There is no need to make a crossbow skin incision. In older children, a mastoid-to-mastoid incision may be preferable, particularly if a wide exposure of one or both cerebellar hemispheres is desired. However, by far the majority of infratentorial tumors in childhood are mid-line in location and the mid-line exposure, which is more quickly and bloodlessly opened and closed, is entirely adequate.

It is our policy to include an occipital burr hole in the operative field routinely even if ventricular estimation is not performed. This burr hole is placed rather low; that is, about 2 to 3 cms. from the mid-line and 2 to 3 cms. above the location of the transverse sinus in case it is necessary to perform a Torkildsen procedure. The lateral ventricle is always tapped through the burr hole to relieve any excessive pressure before the dura of the posterior fossa is incised.

In addition to removing a generous amount of the occipital bone bilaterally, in tumor patients the arch of the atlas is usually removed as well. This is always done if there is herniation of the cerebellar tonsils through the foramen magnum or extension of the tumor itself into the upper cervical canal. In the latter situation, it may be necessary occasionally to remove the laminae of the second or even the third cervical vertebra also. The dura is opened in a cruciate manner over each cerebellar hemisphere. The occi-



Figure 205. Infant on operating table in position for posterior fossa exploration. Endotracheal ether anesthesia is used with the patient's head in a well padded horseshoe shaped cerebellar head rest. Thermocouple in the baby's rectum for constant temperature recording Plastic catheter in right ankle vein for constant infusion of fluids and blood. Stethoscope strapped to the posterior chest wall is used for checking the rate and character of respirations Patient lies on an electric pad to be used if temperature goes down unduly.

pital sinus is divided between ligatures or silver clips. If tapping the lateral ventricle does not reduce the tension of the posterior fossa dura so that it pulsates and becomes somewhat slack, it is wise to make only a nick in the dura over each hemisphere and introduce a ventricular cannula in search of a cyst before the dura is opened widely.

When the dura has been opened, a tumor or cyst may be visible on the surface, or one hemisphere of the cerebellum may be so enlarged or distorted as compared with the other to point to the location of an underlying tumor. If the hemispheres appear normal but separated from one another by a prominent, bulging vermis, the tumor is usually primarily in the midline. When the tumor lies entirely within the fourth ventricle or deep in the mid part of the cerebellum, the only visible deformity may be herniation downward of the cerebellar tonsils. In exploring over the surface of the cerebellum particular care should be taken not to disrupt the veins bridging to the tentorium.

If no tumor is visible on the surface and no distortion of the hemispheres is evident, the tonsils should be separated carefully after opening the arachnoid of the cisterna magna so that the posterior end of the fourth ventricle can be visualized. If this does not disclose tumor, then the vermis

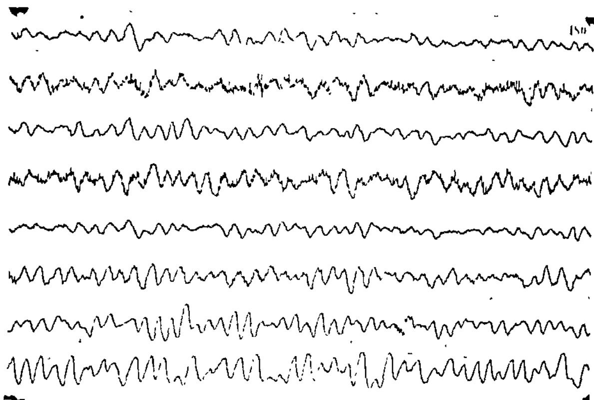


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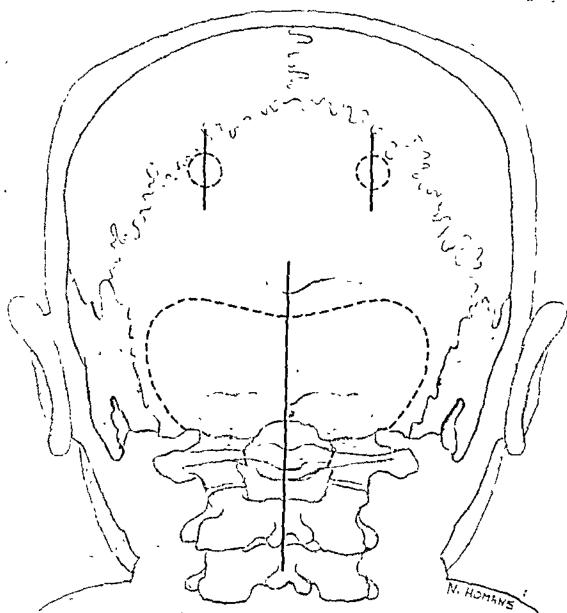


Figure 206. Plan for exposure of posterior fossa in infants and children. Heavy line indicates position of skin incisions. Dotted lines indicate positions of burr holes and extent of sub-occipital craniectomy. This may or may not include the arch of the atlas as necessary.

should be split in the mid-line after cauterization of surface vessels. The operator should never assume a negative posterior fossa exploration until the entire surface of the cerebellum has been examined including both cerebello-pontine angles, until both cerebellar hemispheres have been explored with a ventricular cannula, and the entire fourth ventricle including the posterior end of the aqueduct has been visualized directly, if necessary after splitting the vermis.

The dura is always left open after removal of posterior fossa neoplasms

in children. The muscular and cutaneous layers are closed meticulously in multiple layers with interrupted sutures. Especial care is necessary to achieve a careful closure of the muscles in the mid-line at the point of attachment to the occipital bone; this is always the most vulnerable point in the closure. After the ordinary type of dressing is applied, it is wise in children to add a halter type of restraint by placing long adhesive tape strips from the top of the head down the back in order to prevent sudden acute flexion of the neck during the immediate post-operative period (Figure 207).

An effort is made to employ electrocautery as little as possible in the neighborhood of the fourth ventricle and medulla oblongata. After any surgical procedure in the posterior fossa, complete hemostasis and removal of all devitalized and necrotic material should be accomplished to minimize the formation of post-operative arachnoid adhesions. There should be visible demonstration of free circulation of spinal fluid through the aqueduct and fourth ventricle before closure is performed. If this is impossible due to aqueductal, or some other type of inoperable obstruction, a Torkildsen tube should be placed from one lateral ventricle to the posterior fossa at the same operation (p. 136).

At the time of final delivery or rapid suction-removal of large posterior fossa tumors the patient's vital signs should be watched with great care. Spontaneous respirations may cease or become very irregular for a time and the anesthetist should be prepared to carry the patient through this period on artificial respiration. Whole blood should always be running in

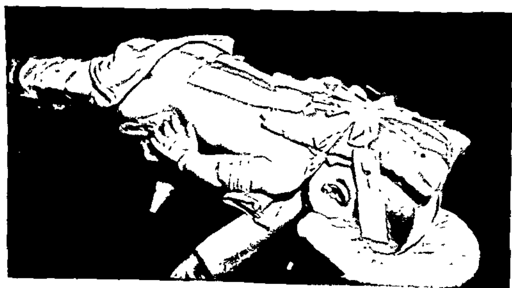


Figure 207. Dressing applied post-operatively following posterior fossa surgery in infants. Sterile gauze flats over the incision are covered by a folded sterile towel and held in place by adhesive tape straps placed on the head, shoulders and back with tapes tied over the sterile towel. Patient is then turned from side to side during the post-operative period.

freely during this phase of the operation, and in young children particularly, an attendant should be free to pump in whole blood rapidly should this prove necessary.

Under ordinary circumstances the entire tumor should be removed in one stage wherever possible. Occasionally, if the child's general condition is poor, removal may well be done in two stages a week or so apart. However, if moribund or very poor risk patients are put on constant ventricular drainage (p. 136) for a few days and improved as far as general condition is concerned, one-stage procedures are usually possible and preferable.

Complications

Post-operative complications in infratentorial tumor surgery in children include: (1) hyperthermia; (2) transient cranial nerve palsies; (3) wound hematomas; (4) aseptic meningitis; (5) gastro-intestinal ulceration and hemorrhage; and (6) false meningocele formation.

Hyperthermia: This is fairly common after removal of lesions within the fourth ventricle or encroaching upon the aqueduct. It is combatted by taking rectal temperatures at regular, frequent intervals post-operatively and starting antipyretic treatment before the temperature gets too high. It is much easier to prevent the temperature from going above 103° F. than it is to bring it down again when it has reached 105° F. Artificial means of cooling include the use of ice-bags, electric fans, alcohol sponges, ice-water enemas, salicylates in large doses by rectum, and parenteral fluids.

Cranial Nerve Paralysis: Because of the location of these lesions, transient palsies of the sixth, seventh, eighth, ninth, and tenth cranial nerves may occur after operation. These are important particularly in so far as swallowing of secretions and vomiting are concerned. Adequate suction should always be available to take care of oral and nasopharyngeal secretions post-operatively. It may be impossible to give fluids by mouth for many days, so that parenteral fluid therapy and gavage feedings for prolonged periods should be employed when necessary.

Wound Hematomas: This occasional complication of all intracranial surgery is especially important in posterior fossa wounds because since the bone is removed and the dura left open, a secondary wound hemorrhage here may produce complete obstruction to spinal fluid circulation with sudden and drastic sequelae. Any post-operative swelling of the wound which is not relieved by lumbar or ventricular drainage of spinal fluid demands re-exploration of the operative incision.

Aseptic Meningitis: After a certain number of removals of cerebellar tumors, when a large empty space is left behind, the post-operative period is characterized by persistent low-grade fever, clinical signs of meningeal irritation, and a spinal fluid pleocytosis of 20 to 500 white blood cells, most

of which at first are polymorphonuclear leukocytes, but soon become almost entirely mononuclear cells. Presumably this is an aseptic meningeal reaction to blood and other breakdown products from the tumor bed which enter the spinal fluid circulation. It usually lasts from one to three weeks.

Gastric Ulceration: One of the most distressing and severe complications of posterior fossa surgery is the development of so-called "Cushing-Rokitansky" ulceration of the stomach or duodenum. This complication is frequently fatal due to massive hemorrhage into the intestinal tract or to perforation and sudden overwhelming peritonitis. It usually appears within two or three days after operation but occasionally as late as a week. Minor degrees of gastric bleeding have been treated successfully with constant alkalinization of the upper intestinal tract, administration of atropine, replacement of blood loss and general supportive measures. There seems to be no way to predict pre-operatively or at the time of operation which patients may develop this critical complication.

False Meningoceles: Because the dura is left open and the bone removed, and because the muscles available for closure are often thinned out markedly, there may be considerable distention of the suboccipital decompression, particularly if post-operative edema causes temporarily some obstruction to the normal circulation of spinal fluid. This results in a collection of spinal fluid within the wound which becomes lined by a smooth, dura-like membrane. Secondary wound revision, with excision of this membrane, resuture of the muscular layers, or reinforcement of the closure with fascial grafts or tantalum mesh may be necessary to obliterate this false meningocele. Such a procedure can be successful only if there is no obstruction to spinal fluid circulation and absorption.

MEDULLOBLASTOMA

The term medulloblastoma was coined by Bailey in 1924[11] to describe a group of gliomas characteristically found in the posterior fossa of children, arising usually in the region of the anterior medullary velum and occupying a mid-line position within the substance of the cerebellum. Since this description and the subsequent extensive review of Cushing in 1929,[43] there has been general agreement concerning the clinical and pathological features of this common intracranial neoplasm.

According to Bailey,[11] the medulloblast is an embryonal cell of an indifferent character derived from the primitive medullary *epithelium* which migrates and divides actively during the normal histogenesis of the brain and may later differentiate into either neuroblast or *spongioblast*, or may remain undifferentiated. "Rests" of such embryonal cells may occur at various places throughout the brain and particularly *in the region of the*

roof of the fourth ventricle. It seems probable that the rapidly growing undifferentiated medulloblastoma of early childhood may well arise from such persistent groups of embryonal cells.

Clincal Features

Among the posterior fossa tumors of infancy and early childhood, this lesion is the most frequent. In our series, the diagnosis of medulloblastoma has been verified histologically in 68 patients under 12 years of age. Although this tumor occurs in adolescents and also occasionally in adults,[186] the peak of frequency is in the first decade of life. In our series the age of patients at the time of hospital admission varied from one month to 11⁹⁄₁₂ years, with a striking peak at three years, as shown in Figure 208. There were 41 males and 27 females.

Of the posterior fossa neoplasms in childhood, the medulloblastoma is the most rapidly growing. All of the symptoms and signs described in the general discussion of infratentorial tumors (p. 225) are seen in their most

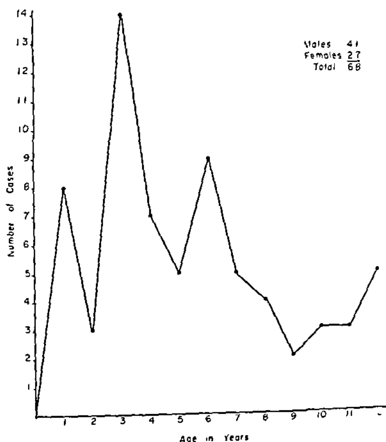


Figure 208. Age at time of hospitalization of 68 patients under 12 years of age with medulloblastomas in the posterior fossa.

extreme form and progress in their most relentless fashion with this lesion. Even more than with astrocytomas, the early symptoms and signs relate to increased intracranial pressure rather than to the location of the tumor itself. Vomiting, headache and unsteadiness of gait, accompanied subsequently by visual complaints, papilledema, enlargement of the head, increasing weakness and possibly tonic seizures proceed according to an almost uniform pattern. Among our patients, the duration of symptoms varied from 10 days to 14 months, the average being four to six weeks (*Figure 209*).

The average child with a medulloblastoma gives the general appearance of much more severe illness than does the child with less malignant varieties of posterior fossa tumor. *It is well to stress again here, however, that there are no reliable criteria of the tumor type previous to operative visualization.*

Among our patients, 10 showed normal eye-grounds at the time of admission and 55 showed papilledema. The others were equivocal or not recorded. Roentgenograms of the skull were interpreted as normal in 20 patients; in 48 there was separation of the cranial sutures or other x-ray

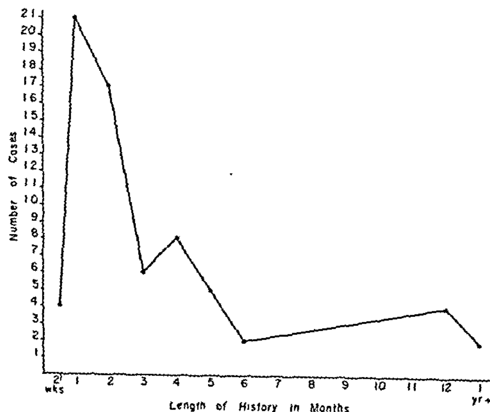


Figure 209. The length of history in children with cerebellar medulloblastoma. Compare with Figure 215 showing similar data in children with cerebellar astrocytoma.

evidence of increased intracranial pressure. Ventriculograms were performed in 34 patients and invariably showed symmetrical dilatation of the lateral and third ventricles. Air in the fourth ventricle is seldom seen in patients with medulloblastoma, because the caudal aperture of the aqueduct is usually blocked by tumor (Figure 210). Thirty-four patients were operated upon on the basis of clinical findings and ventricular estimation alone.

Morbid Anatomy

In almost every instance the medulloblastoma of childhood is principally located in the mid-line. It is closely attached to the region of the anterior medullary velum in the roof of the fourth ventricle. It may extend for a variable distance toward the dorsal surface of the vermis, and when it does so, produces widening of the folia of the vermis, prominence of the entire vermis, separation of the cerebellar hemispheres and herniation downward of the cerebellar tonsils through the foramen magnum. The tumor may extend more into one cerebellar hemisphere than another, producing surface asymmetry, but this is unusual. It invariably grows into the lumen of the fourth ventricle and may completely fill it, extending well forward into



Figure 210 Lateral ventriculogram of seven year old child with posterior fossa medulloblastoma There is enlargement of the lateral and third ventricles and upper aqueduct The block in the mid portion of the aqueduct is easily seen (arrow)

the aqueduct and laterally into the lateral recesses. Tumor frequently protrudes from the caudal end of the fourth ventricle and becomes visible overlying the region of the calamus scriptorius (Figure 211). Such a protrusion may be fairly extensive and easily seen within the cisterna magna as soon as the dura is opened, or it may protrude as a small nubbin from under the margin of the prominent vermis after separation of the tonsils. Tumor may also grow out through the foramina of Luschka laterally.



Figure 211 Large medulloblastoma in a two year old child protruding from the fourth ventricle, filling the cisterna magna and displacing the cerebellar hemispheres laterally.

One of the striking features of the medulloblastoma is its tendency to "infect" the subarachnoid pathways; that is, to seed to distant points in the meninges. Separate small nodules of tumor may be seen occasionally over the surface of one cerebellar hemisphere, in the cerebello-pontine angle, or over the upper cervical cord. Prior to death widespread seeding through-

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out the spinal meninges, the ventricular system and particularly the basilar cisternae is common.

The medulloblastoma is a reddish gray or purplish, relatively vascular tumor whose surface when exposed is usually irregular and crossed by numerous discrete blood vessels. The tumor is soft and friable and usually readily "suckable." There is no clear line of demarcation from the surrounding cerebellum although a false plane of cleavage may often seem to develop. It does not invade the ependyma lining the floor of the fourth ventricle. These lesions are rarely cystic, but occasionally one or more small cavities containing several cubic centimeters of yellowish fluid are present.

Microscopically, the medulloblastoma is an extremely cellular tumor consisting almost entirely of masses of small, round or slightly oval cells which are arranged in no definite pattern other than a frequent tendency to acinar or pseudo-rosette formation (Figure 212). The nuclei are prominent, the cytoplasm scanty, and fibrils are not ordinarily demonstrable. There are many mitotic figures. There is little stroma and microscopically this appears largely confined to the vascular channels. With heavy metal stains, occasional spongioblasts and neuroblasts may be demonstrated. In some of these tumors, an unusual number of neuroblasts, that is, cells with large clear nuclei and a prominent nucleolus, may be present associated with more than the ordinary amount of connective tissue stroma. While some pathologists have designated these lesions neuroblastomas, we have followed the practice of including them among the medulloblastomas.

Treatment

The treatment of medulloblastomas of the posterior fossa is certainly one of the darkest chapters in pediatric neurosurgery.[93] To our knowledge no patient with this lesion has ever been cured, and it seems unlikely that one ever will be with our present therapeutic methods. Complete surgical removal is evidently impossible, since no matter how thorough an excision has been performed and how free from gross evidence of tumor the posterior fossa appears, the lesion always recurs and eventually causes death. Unfortunately, there is no way to identify this tumor certainly before operation, as has previously been stressed.

The medulloblastoma is extremely sensitive to x-radiation and to date this constitutes the most effective form of treatment. Primary x-ray treatment on the assumption of clinical diagnosis, however, is never justified in our belief for several reasons:

(1) There is almost an even chance statistically that the tumor is an astrocytoma and therefore subject to the possibility of complete cure by surgical removal.

(2) X-ray treatment without suboccipital decompression may cause

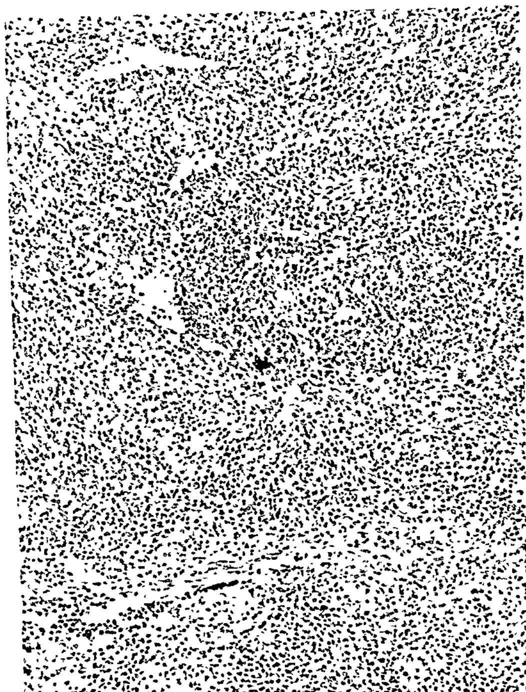


Figure 212. TBE x 200 Photomicrograph from mid-line soft vascular medulloblastoma attached to the roof of the fourth ventricle of a 1 11/12 year old infant. The tumor consists of masses of densely staining small cells with very scanty stroma. Mitotic figures are frequent and many anaplastic cell forms are seen. Occasionally the tumor cells seem to be oriented radially around blood vessels.

temporary increase in the size of a medulloblastoma with resulting further bulbar compression of serious degree.

(3) Irreversible loss of vision often occurs very rapidly secondary to papilledema in childhood and to have this occur during a so-called "trial" period of x-ray therapy is an unnecessary tragedy.

(4) The most satisfactory immediate relief of the distressing symptoms of increased intracranial pressure is achieved by surgical re-establishment of spinal fluid circulation.

Operation

The surgical approach to the posterior fossa has already been described (p. 235). The arch of the atlas is removed only if the cerebellar tonsils or tumor proper protrude below the foramen magnum. It is our custom never to rely entirely upon the gross appearance of the lesion but to obtain a biopsy for histological study in every patient. It is then our purpose not to attempt a total removal of medulloblastomas but to remove sufficient tumor to re-establish normal spinal fluid circulation. Since the great mass of the tumor is usually located within the lumen of the fourth ventricle, this ordinarily amounts to a subtotal removal of the lesion.

Although there is no clear line of demarcation from the cerebellum, *fortunately there is ordinarily no invasion of the floor of the fourth ventricle* or the walls of the aqueduct. It is usually possible to expose the posterior end of the fourth ventricle and elevate the tumor enough to place moist cotton strips between it and the floor of the ventricle. When this has been done and the tumor well exposed by splitting the vermis and developing its lateral margins, the bulk of the tumor is removed quickly by suction. Surface vessels may be coagulated, but there is little point in attempting to control the bleeding within the tumor itself until the great majority of it is removed. When tumor overlying or extending into the lumen of the aqueduct is dislodged there is an immediate gush of clear fluid from the third ventricle. The aqueduct must be unblocked to insure adequate postoperative spinal fluid circulation. If it cannot be exposed or unblocked, a Torkildsen tube should be placed to by-pass this obstruction (p. 136). When free circulation of spinal fluid has been established and complete hemostasis achieved, the dura is left open and the rest of the wound closed in layers in the usual manner.

X-ray Treatment

It is our feeling that the type of surgery outlined above followed by intensive deep x-ray therapy to the entire cerebrospinal axis offers the child with medulloblastoma a maximum interval of comfortable life, free of increased intracranial pressure and symptoms of cerebellar or brain stem compression, followed by a rapid terminal illness. Since cure by present methods has proved impossible, efforts have been directed toward prolongation, not necessarily of life, but of the symptom-free period of survival.

X-ray therapy may be started ordinarily as soon as the patient has stabilized after surgery, that is, within five to 10 days. Barbiturate sedation,

or even rectal avertin analgesia may be necessary in restless young children for each treatment. Therapy is first directed to the mid-line of the posterior fossa, a total tumor dose of about 3,000 R being administered in an average of 15 to 20 treatments through three portals. Following this an additional tumor dose of 2,000 to 3,000 R is directed fractionally to the rest of the cerebrospinal axis. Such a course of therapy may be repeated after three to six months if desired. However, if symptoms recur within this interval, deterioration and death are apt to ensue rapidly regardless of treatment. Complications of x-ray treatment include vomiting, leucopenia, and rarely acute cerebral edema.

Results

In our series of 68 children under 12 with cerebellar medulloblastomas there have been 18 post-operative deaths; this includes all deaths during the first month after operation. Forty-five patients received deep x-ray therapy after surgical verification of the tumor type and re-establishment of spinal fluid circulation. The survival times in this group of patients are shown in Figure 213. An occasional long survival has been reported in a patient with medulloblastoma,[91, 157] but to our knowledge this has never occurred when the original symptoms began during early childhood.

Cerebellar Medulloblastoma, Time of Death Postoperative

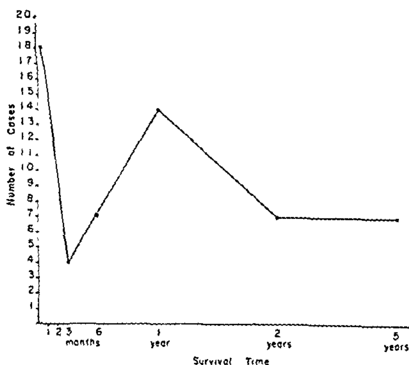


Figure 213. Survival period of patients after surgical confirmation of cerebellar medulloblastoma

the mid-line but extended into one hemisphere as well for some distance, and 19 were located entirely within one cerebellar hemisphere (Figure 216). Perhaps the outstanding characteristic of this tumor is the tendency to cyst formation. These cysts may become very large and may lie in any relation to the solid portion of the tumor which is often a relatively small mass, or nodule, limited to one particular part of the cyst. The greater part of the cyst wall may be thin and may either be present on the surface (Figure 217) or be entirely surrounded by cerebellar tissue. It is uncertain under such circumstances whether there are tumor cells in the entire cyst wall. It is necessary to remove the entire tumor nodule and adjacent cyst wall for surgical cure, but in some instances at least cure may follow when portions of the cyst wall are left behind. In many instances there is no discrete nodule, but the entire cyst is surrounded by a layer of solid tumor.



Figure 216 Operative exposure of a large solid astrocytoma of the right cerebellar hemisphere. Note enlargement of this hemisphere, widening of the folia, increased vascularity and translucency of the overlying pia-arachnoid. There was also herniation of the cerebellar tonsil of this hemisphere through the foramen magnum.

The fluid within astrocytoma cysts is usually clear or slightly translucent, it varies from pale yellow to an amber color, and may have such a high protein content that it gels within a few moments after removal. In our series, 31 of the cerebellar astrocytomas were cystic, though in some cases the cysts were rather small so that the greater part of the mass was actually solid tumor. Twenty-five of the tumors were entirely solid (Figure 218).

The solid portion of a cerebellar astrocytoma is ordinarily a pinkish or yellowish gray color, is tough and rubbery in consistency so that it is not easily "suckable," is comparatively avascular, and is fairly well demarcated from the surrounding cerebellum. There is a thin layer of compressed, often necrotic brain tissue immediately adjacent to the tumor, and as far as possible surgical extirpation should be carried out in this layer. Small vessels bridging to the capsule of the tumor from the cerebellum are readily controlled if the exposure is adequate.

Unfortunately, the solid, mid-line astrocytomas have a great tendency to extend into the region of the cerebellar nuclei and peduncles and upward along the Aqueduct of Sylvius. When this is true, or when extension occurs



Figure 217. Cystic astrocytoma of the cerebellum in a 13 months old infant exposed at operation. There was a tumor nodule in the mid-line with a huge cyst occupying most of the left half of the posterior fossa. Complete removal was carried out and patient has been asymptomatic for two and a half years since operation.

actually into the brain stem, complete removal may become impossible, or at least inadvisable. Solid tumor may fill the fourth ventricle and may extend either upward through the lumen of the dilated aqueduct into the third ventricle or downward into the cervical spinal canal. Obviously, the surgical results with cystic lesions of the hemispheres are vastly superior to those with the primarily solid mid-line tumors.

Microscopically, these tumors consist largely of a fairly uniform feltwork containing evenly distributed astrocytes (Figure 219). The cellularity and degree of edema vary considerably from one tumor to another but the cell type remains fairly constant (Figure 220). No attempt is ordinarily made to differentiate fibrillary and protoplasmic varieties among the cerebellar astrocytomas. The cytoplasm of the astrocytes is usually scanty; the nuclei are oval and eccentrically placed. Mitotic figures are rarely seen. The fibrils within the processes are well brought out with phosphotungstic acid hematoxylin stain. Blood vessels are uncommon.

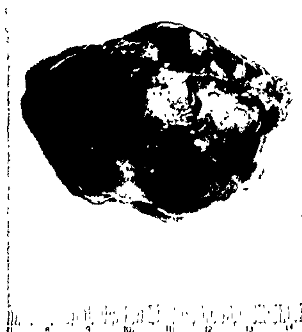


Figure 218. Solid astrocytoma removed intact from the right cerebellar hemisphere of a five year old child. Negative neurological examination and no evidence of increased pressure three and a half years after operation.

Treatment and Results

The treatment of cerebellar astrocytomas is entirely surgical. X-ray therapy does not alter the growth of these tumors, nor is there evidence that their growth is influenced by any chemical agent. The surgical approach to the posterior fossa has already been described (p. 235). Simple evacuation of an astrocytomatous cyst may relieve symptoms completely for a long time; however, the cyst will always refill and symptoms recur un-

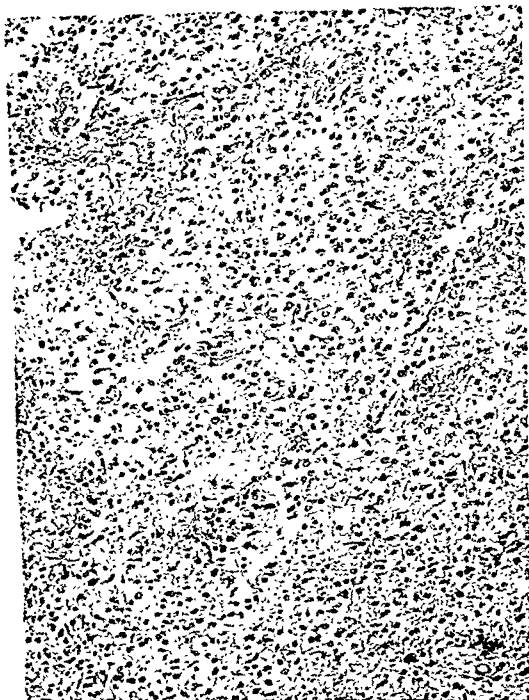


Figure 219 PTAH x 200 Photomicrograph of section from large solid fibrillary astrocytoma removed from the right cerebellar hemisphere of a five year old girl. Note the diffuse arrangement of astrocytes in a very loose stroma with no perivascular arrangement The astrocytes have many finely branching processes. No mitoses are seen. Same tumor shown in Figure 218.

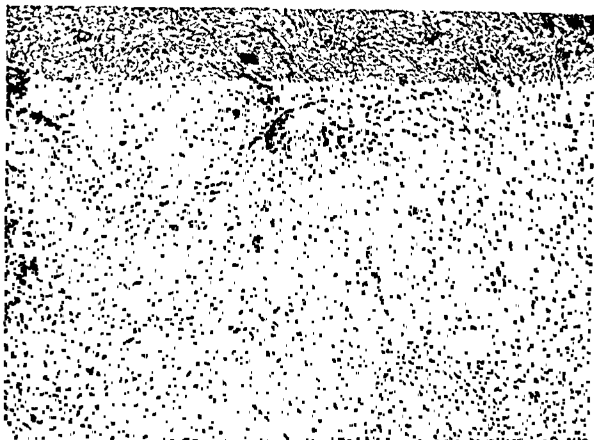


Figure 220. PTAH x 200 Photomicrograph of astrocytoma removed from the cerebellar vermis and fourth ventricle of a 4 8/12 year old boy. The tumor consists of cellular masses interspersed with many blood vessels. There is considerable necrosis and interstitial hemorrhage. Multipolar astrocytes are scattered diffusely without any tendency to perivascular arrangement No mitoses are seen The tumor appears moderately edematous.

In this series of 56 cerebellar astrocytomas treated surgically, there were nine deaths during the post-operative period. This mortality is higher than some reported series because it includes only patients under 12 years of age, and the surgical results have been less satisfactory the younger the patients. A total of 47 patients are alive and free of symptoms of increased intracranial pressure from three months to 20 years.

EPENDYMOMA

Next in frequency to astrocytoma and medulloblastoma among infratentorial neoplasms of childhood is the ependymoma. Although this tumor is often of relatively low-grade malignancy with respect to its rate of growth and its invasive character, its common close attachment to the floor of the fourth ventricle usually makes its complete removal surgically impossible and the hazards of even partial removal unusually great.

The clinical picture may be entirely that of increased intracranial pressure as already outlined. However, because this lesion so often extends downward into the upper cervical spinal canal, stiffness and spasm of

the neck and shoulder muscles, limitation of neck motions and suboccipital and upper cervical spine tenderness are also commonly found.

In our series of 185 posterior fossa tumors, there were 19 ependymomas. These patients ranged in age from two months to seven and a half years. There were 13 males and six females. The average duration of symptoms was about two months.

At the time of hospital admission, 15 of our patients showed papilledema. On x-ray examination, separation of the cranial sutures was present in 13 and there was no x-ray evidence of increased intracranial pressure in four. Two patients showed calcification within the tumor in the plain x-ray films (Figure 222).



Figure 221. Seventeen year old girl leading normal life 14½ years after removal of cerebellar astrocytoma. Negative neurological examination.

Morbid Anatomy

This tumor characteristically occupies the caudal portion of the fourth ventricle and apparently arises from ependyma in this region. It may fill the entire fourth ventricle, extend into the lumen of the aqueduct and protrude inferiorly well down into the upper cervical canal (Figure 223). However, its point of attachment is almost always the floor of the fourth ventricle in the region of the calamus scriptorius. It is usually visible projecting into the cisterna magna as soon as the dura has been opened at operation.

More rarely this tumor apparently arises in the region of the apex of one lateral recess of the fourth ventricle. When this occurs, it occupies the cerebello-pontine angle and may grow to fill a large part of one half of the posterior fossa without encroaching directly into the lumen of the fourth ventricle itself. The cerebellum is displaced much more than it is invaded.

Posterior fossa ependymomas vary a great deal in their gross appearance, as they apparently do in their rate of growth. Usually they are firmer and less vascular than the medulloblastomas. The tumor is often variegated on the surface, with yellowish, pink and gray areas intermingled. It is less friable than the medulloblastoma because, although extremely cellular, there is more stroma. Flecks of calcium may occasionally be seen or felt

within the tumor substance. There is usually a fairly well defined layer of cleavage between the tumor and the surrounding tissues everywhere except at the point of its ependymal attachment. Cyst formation is uncommon.

Microscopically the ependymoma characteristically shows an abundant stroma. There are many vascular channels around which are grouped polygonal or slightly elongated cells in a radial pattern (Figure 224). The frequent appearance of clear areas adjacent to these vascular channels is due to the presence of many elongated processes the cell bodies of which are more peripherally located. Away from the vessels the cells are arranged in a more homogeneous mosaic (Figure 225). Mitotic figures are uncommon. Small, stainable bodies within the cytoplasm, known as blepharoplasts, are often seen in the elongated neoplastic cells.



Figure 222. Basal roentgenogram showing calcification in an ependymoma of the fourth ventricle.

Treatment and Results

As already mentioned, it is ordinarily impossible to remove ependymomas of the fourth ventricle completely because of their inseparability from vital areas. Since the tumor is generally well demarcated and is also relatively avascular, it can be removed piecemeal and by suction with comparative ease until the floor of the fourth ventricle is approached. A thin layer of tumor must then be left behind to avoid damage to the cranial nerve nuclei, restiform bodies, and nuclei of the dorsal columns of the cord. Electro-dissection near the floor of the fourth ventricle should be avoided.

This type of surgical excision should re-establish spinal fluid circulation and therefore provide immediate relief of symptoms of increased intracranial pressure. It may be necessary to perform laminectomy of C_1, C_2 and occasionally even C_3 to remove intraspinal extension of this lesion. Theoretically, complete excision of an ependymoma in the lateral recess and cerebello-pontine region without undesirable loss of function should be possible.

In this clinic post-operative deep x-ray therapy is directed to the posterior fossa in all patients with ependymoma A tumor dose of about 3,000 R is administered through three portals. The rest of the cerebrospinal axis is not ordinarily treated.



Figure 223. Post-mortem sections through the fourth ventricle showing complete obliteration of the lumen and displacement of the cerebellum by ependymoma.

Until recent years the immediate post-operative mortality with these lesions was extremely high. In the last four years there has been one post-operative death among seven children with fourth ventricle ependymomas. Five patients of this entire group of 19 patients are still alive from eight months to 10 years after operation and x-ray treatment.

GLIOMAS OF THE BRAIN STEM

It is unfortunate that one of the more common locations of gliomas in childhood is within the brain stem; that is, in the portion of the brain between the hypothalamus and the upper level of the cervical spinal cord.



Figure 224. TBE x 200 Photomicrograph of section from solid ependymoma removed from the fourth ventricle and upper cervical spinal canal of a five and a half year old boy. Note characteristic arrangement of pear-shaped glial cells around the blood vessels with elongated processes extending toward the vessel wall. This gives the appearance of a band of collagenous tissue immediately adjacent to the vessel. Elsewhere the tumor is markedly edematous with a loose arrangement of the tumor cells. Occasional mitoses are seen.

These infiltrative gliomas are particularly frequent in the pons and upper medulla oblongata. Regardless of specific histology, they must all be classified as malignant tumors since their location in itself renders them inoperable. Actually, they have usually proved to be astrocytomas, spongioblastomas, or mixed gliomas, portions of which often resemble closely the glioblastoma multiforme seen so commonly in the cerebral hemispheres of adults.[23]

Because of the absence of symptoms and signs of increased intracranial pressure, often until death, the diagnosis of brain stem glioma is frequently long delayed. In this respect these tumors differ widely from the commoner gliomas of the cerebellum and fourth ventricle in childhood where there may be no neurological deficit but only increased pressure until late in the course of the disease Brain stem tumors are mistaken for cerebral palsy, various types of encephalomyelitis, bulbar poliomyelitis, cerebral vascular disorders, and tuberculous and other forms of chronic meningitis.

The incidence of brain stem gliomas cannot be stated with the same

accuracy as other posterior fossa neoplasms since frequently a histological diagnosis is lacking. Many patients with a satisfactory clinical or x-ray diagnosis are not explored surgically because of the hopeless prognosis and high mortality of operation. Some of these patients, therefore, perhaps after a course of x-ray therapy, die at home and autopsy is not performed.

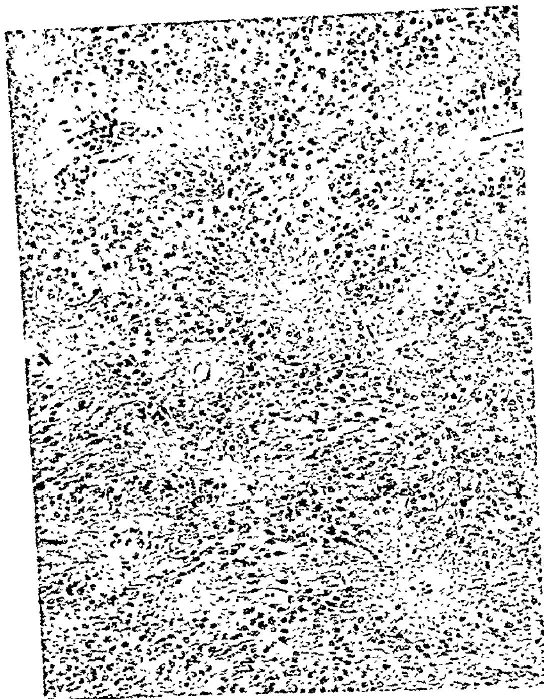


Figure 225 PTAH x 200 Photomicrograph of ependymoma of the fourth ventricle in 2 3/12 year old boy Note arrangement of blood vessels surrounded by a thick layer of collagen then a layer of cells giving a rosette appearance. In between this paravascular configuration the cells are loosely arranged in a fibrillary stroma.

In our series, the clinical diagnosis of brain stem tumor has been made 30 times in children under 12 years of age. Confirmation was obtained at operation or autopsy in 23 patients; in the other seven, the diagnosis was based on clinical and ventriculographic examination alone. The age and sex incidence of these lesions are shown in Figure 226.

Clinical Features

In the brain stem are located most of the cranial nerve nuclei and through it pass all of the ascending and descending connections of the cerebrum with the spinal cord and cerebellum. It is natural to expect, therefore, that a wide variety of neurological symptoms and signs may result from a lesion within its confines. As already suggested, perhaps the most characteristic feature of these tumors is their progression to advanced stages without development of increased intracranial pressure. Headache, papilledema, enlargement of the head, and separation of the cranial sutures are usually absent. The length of the history is usually from one to five months (Figure 227).

GLIOMAS OF THE BRAIN STEM

AGE INCIDENCE

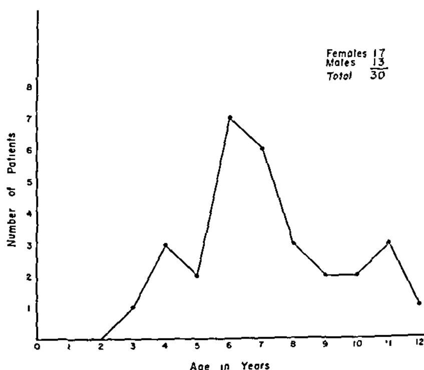


Figure 226. Age and sex distribution of children with gliomas of the brain stem Almost two-thirds of these patients were between five and eight years of age

GLIOMAS OF THE BRAIN STEM

DURATION OF SYMPTOMS

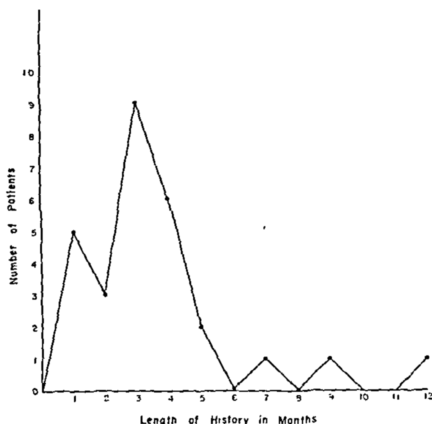


Figure 227. Length of history in 30 children with gliomas of the brain stem. In general the history is shorter than in patients with other posterior fossa tumors.

Figure 228. Seven year old girl with glioma of the brain stem. Note extensor rigidity of all extremities, bilateral facial palsy and profound coma. Patient earlier demonstrated bilateral sixth nerve palsies and unilateral eighth and ninth cranial nerve palsies together with spontaneous nystagmus. There was no increase in intracranial pressure.

Symptoms and Signs: An almost pathognomonic syndrome of this lesion consists in the presence of multiple, bilateral cranial nerve abnormalities in combination with pyramidal signs and truncal ataxia in the absence of increased intracranial pressure. For all practical purposes there are no other lesions which will give this clinical picture in a young child. The cranial nerve nuclei involved are the lower group, principally five through nine, and occasionally twelve.

The commonest cranial nerve involved is the sixth (22 times). Often the weakness of lateral gaze is bilateral. The seventh nerve is also commonly involved (19 times); facial diplegia has been seen on several occasions (Figure 228). Facial weakness is ordinarily of the peripheral type since the nucleus itself is concerned. Fifth nerve involvement is best established in young children by testing the corneal reflexes (7 patients). Eighth nerve function is difficult to evaluate till late in the disease, although diminished hearing has sometimes been detected early in older children (5 patients). Ninth and tenth nerve dysfunction is manifested by difficulty with swallowing and articulation and is demonstrated by palatal weakness, pharyngeal anesthesia and by abnormality of vocal cord movement (15 patients). Frequent protracted vomiting in the absence of increased pressure is undoubtedly due to direct involvement of vagal nuclei. Seventeen out of 28 patients had a history of vomiting but only seven showed papilledema.



Figure 229. Occiput-up lateral encephalogram showing marked displacement of the aqueduct and normal-sized fourth ventricle due to an infiltrating glioma of the brain stem.

The *cortico-spinal tracts are almost always implicated early* in the course of these lesions. It is therefore usual to find hyperactive deep tendon reflexes, particularly in the lower extremities, with ankle clonus and extensor response to plantar stimulation (25 out of 28 patients). *This is in contradistinction to patients with cerebellar gliomas, where the deep tendon reflexes are ordinarily hypoactive.* When weakness develops, it is spastic rather than flaccid This may be in the form of a hemiplegia, paraplegia or quadriplegia. As the lesion progresses a decerebrate type of spastic quadriplegia ensues (Figure 228).

Because of encroachment on the cerebello-dentate-rubro-thalamic tracts

presumably, gait disturbance is an early complaint (26 out of 28 patients). The patient is unsteady in the Romberg position, walks with a wide base and tends to weave from side to side and fall frequently. Nystagmus and ataxia of the extremities themselves are less common.

When tumor invades or compresses the higher levels of the brain stem pathological somnolence occurs. Such patients, diligently nursed and nourished, may continue sleeping peacefully for many weeks or even months.

Lumbar Puncture: Since increased intracranial pressure is not present usually, lumbar puncture can be carried out with little or no danger. Spinal fluid pressure is ordinarily not elevated (only three out of 19 patients tapped). There may be no abnormal cells or a pleocytosis up to 20 to 50 white blood cells. The total protein may be normal or occasionally slightly elevated. If markedly elevated, it means that the tumor has reached the surface and is encroaching upon the surface subarachnoid pathways.



Figure 230. Occiput-up lateral encephalogram of nine year old boy with marked ataxia, multiple bilateral cranial nerve abnormalities and no evidence of increased intracranial pressure. Note marked posterior displacement of the fourth ventricle with convex bulging of the floor into the ventricular lumen. There is also flattening and distortion of the shadow produced by air in the pontine cistern (arrow).

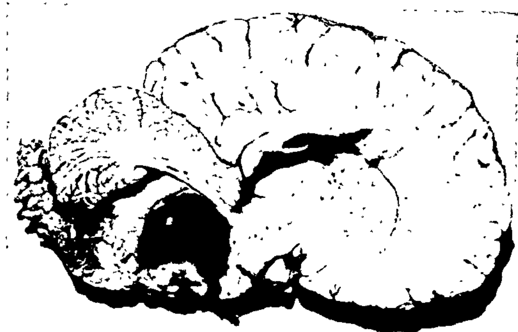


Figure 233. Mid-sagittal post-mortem section showing hemorrhage into a large brain stem tumor. This tumor presented at operation in the fourth ventricle but extended diffusely through the medilla and pons. Patient did not recover consciousness following exploration and biopsy.

by abnormal glial tissue which usually does not obstruct the aqueduct or the lumen of the fourth ventricle (Figure 231). Terminally it often obliterates the pontine and lateral cisternae both by invasion and by compressing them against the floor of the skull and the incisura of the tentorium. The floor of the fourth ventricle is elevated, widened and flattened. This distortion may be all that is visualized if these patients are subjected to posterior fossa exploration. Because of this, biopsy is ordinarily not possible. If tumor is seen in the cerebello-pontine angle projecting from the lateral aspect of the pons, it may be biopsied safely here, though there is little practical purpose in so doing.

Microscopically, in our experience most of these tumors are probably best classified as mixed gliomas, showing areas of astrocytoma, of spongioblastoma polare (bundles of bipolar cells with oval nuclei and delicate, elongated processes), and of glioblastoma multiforme with typical palisading of cells, necrosis, pleomorphism, numerous mitoses and adventitial proliferation (Figure 232).

Treatment

There is need for little comment here since the location of these tumors in itself obviates the possibility of surgical removal. Exploration for confirmation of the diagnosis should be avoided if possible since such surgery is useless (Figure 233). Because there is ordinarily no interruption of spinal

fluid circulation, decompressive or spinal fluid shunting procedures are not indicated. Twenty-one patients in this series were explored. In only five was it possible to obtain a positive biopsy. In recent years, surgery has been avoided when satisfactory clinical and air studies were at hand.

To our knowledge x-ray therapy has never cured a brain stem glioma. The histology of these lesions is not such as to suggest a favorable response to this treatment. However, it is an impression that these unfortunate children often die more comfortably and with occasionally a substantial symptom-free interval after a course of deep x-ray therapy to the brain stem. A tumor dose of about 3,000 R has been used in this clinic.

The average survival time after onset of symptoms and signs has been less than one year, regardless of treatment.

Chapter III

Gliomas of the Cerebral Hemispheres

PUBLISHED REPORTS of gliomas occurring in the cerebral hemispheres in infancy and childhood have been confined largely to descriptions of isolated or unusual cases, very often based solely on pathological studies. Because of the numerical preponderance of gliomas below the tentorium in the pediatric age group, the incidence, symptomatic, radiological, surgical and pathological features of these lesions in the cerebrum have not often been set forward. Ten per cent of the tumors in childhood described by Bailey, Buchanan and Bucy [10] involved the cerebrum, but only three per cent were gliomas. Krayenbühl and Weber [114] reported 62 tumors in children under 17, of which 14 involved the cerebral hemispheres and apparently 10 were gliomas. Keith, Craig, and Kernohan [108] found 49 gliomas among 146 supratentorial tumors in children. Sixteen out of 87 tumors reported by Smith and Fincher [184] involved the cerebrum, and Walker's [200] report of 100 tumors in childhood included nine cerebral gliomas.

CEREBRAL GLIOMAS IN CHILDHOOD



Figure 234 Age and sex distribution of children with gliomas of the cerebral hemispheres

In this clinic, 42 gliomas of the cerebrum were found among a series of 313 consecutive intracranial tumors in children under 12 years of age. From these various series, it is evident, therefore, that the incidence of gliomas of the cerebral hemispheres is about 10 to 14 per cent of all intracranial tumors in childhood. Gliomas arising in the parapituitary region or located primarily within the third ventricle are not included in this group. In our series, except for a peak during the second year of life compensated for by an absence of cases in the following year, the incidence of gliomas year by year throughout early childhood has been essentially constant (Figure 234). More than two-thirds of the group were male; 29 boys and 13 girls.

All of the tumors in this group have been confirmed by histological studies of surgical or post-mortem material. Almost 40 per cent were astrocytomas and another 20 per cent proved to be ependymomas. The balance were scattered among seven other histological types (Figure 235). The striking feature of these figures is the rarity of glioblastoma multiforme in childhood as compared with adult patients.

Symptoms and Signs

Reliable focal signs of cerebral lesions are meagre in small children for reasons which are, for the most part, fairly obvious. The child is unable or unwilling to volunteer complaints and to cooperate accurately in specific tests. Right or left handedness may not have become apparent. Such im-

CEREBRAL GLIOMAS IN CHILDHOOD

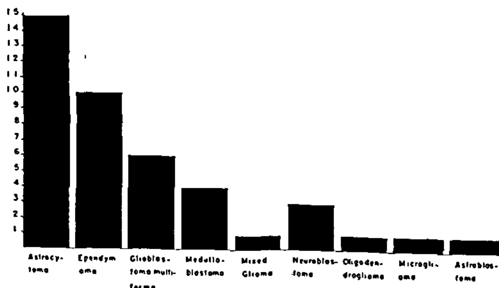


Figure 235. Incidence of various types of gliomas of the cerebral hemisphere seen in children.

portant manifestations of disturbed cortical function as visual field defects, loss of recent memory, astereognosis, aphasia, apraxia, impairment of smell, taste or hearing may be all impossible of reliable evaluation. Even more difficult to determine is the significance of behavior changes during early childhood. Because of this scarcity of reliable symptoms and signs, it is necessary to adopt a high index of suspicion with respect to minor complaints and minimal neurological abnormalities in early life and to pursue investigation freely to the point of such supplementary diagnostic procedures as x-ray examination, spinal fluid studies, electroencephalography, arteriography and ventricular air studies.

Probably the most important primary complaint in these children is one which cannot be defined accurately or properly included in any list of symptoms and signs. It consists in an alteration in the child's disposition, often extremely difficult for the parents or examiner to describe. Listlessness, indifference to surroundings and friends, suppression of the normal enthusiasms of childhood, a difference in emotional response to competitive situations at home or school, the desire to come into the house and lie down frequently, mope quietly or cling to a parent, an unwillingness to cooperate as usual, are all complaints that fit into this category. In retrospect, such ill-defined disturbances of attitude have almost always been present longer than the definitely abnormal symptoms which led to medical investigation.

In this group of patients, headache proved to be a common, and vomiting a rare initial complaint. This is in marked contrast to the more frequent subtentorial neoplasms of childhood where vomiting is a much commoner symptom. The first evidence of focal cerebral involvement was usually motor weakness, most often noted by the child's family as a gait disturbance or a failure to use one hand as well as the other.

Convulsive seizures constituted a primary complaint in only one-third of children with cerebral gliomas. They have not, therefore, been of any great diagnostic or localizing value. In this age group convulsions are prone to occur in a variety of temporary febrile and metabolic disorders in which there are no known structural changes in the nervous system. Conversely, when seizures are associated with local cerebral lesions in children they are nevertheless apt to spread quickly with little clinical suggestion of the epileptogenic site. Rarely in children can one obtain any history of sensory aura. Probably the only type of convulsion that is of value clinically as far as cortical localization is concerned, therefore, is that limited throughout to one side of the face or body or followed by transient hemiparesis.

Physical signs may be almost entirely lacking in these patients until late in the course of the disease. Motor weakness or spasticity may occasionally be demonstrated on examination before it has actually become one

of the patient's complaints. Hyperactivity of the deep tendon reflexes on one side as compared to the other, or the presence of a unilateral extensor plantar response, especially when these appear or become more marked during a period of observation, always point toward a supratentorial lesion.

There is a common impression that papilledema is seen in childhood only with rapidly growing infratentorial neoplasms which abruptly block the circulation of spinal fluid. Although extreme degrees of papilledema accompanied by fresh hemorrhages were not often found in this series, two-thirds of the group demonstrated measurable papilledama at the time of hospital admission.

In our experience the lag between first appearance of symptoms and final establishment of the correct diagnosis has often been a particularly long one among young patients with cerebral gliomas. Primary faulty diagnoses have most often been poliomyelitis (Figure 236), cerebral palsy, birth injury and epilepsy. It is therefore re-emphasized that in this age group, objective diagnostic procedures should be employed freely for minimal indications.

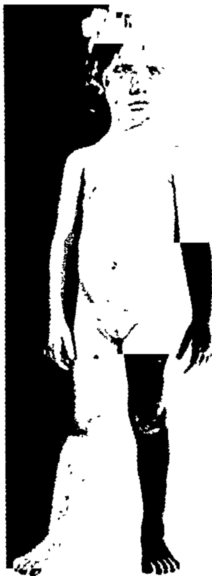


Figure 236. Three and five-twelfths year old girl admitted to the hospital with a question of poliomyelitis because of weakness of the right arm. In addition there was found to be weakness in the right leg and face with early papilledema. There was no history of headache, vomiting or visual disturbance. The patient had a huge cystic fronto-parietal ependymoma of the left hemisphere (Figure 244).

Electroencephalography

The electroencephalogram has provided an invaluable objective means of screening children in whom clinical evaluation of convulsive seizures has done little to identify the etiology or to localize the epileptogenic site. Certainly all children who show localized foci of abnormal electrical discharge, whether bursts of spikes or spike-and-wave complexes (Figure 237), or simply random spikes or groups of slow waves (Figure 238), in association with clinical seizures, headache, or behavior abnormalities should have investigation pursued to the point of ventricular

air studies or arteriography. The commonest electroencephalographic abnormality noted in the presence of cerebral neoplasms in children who have had clinical seizures has been the occurrence of high amplitude slow (two to three per second) waves limited to one or more leads (Figure 239).

Perhaps one of the most frequent errors in the management of intracranial tumors in childhood is to mistake a hemisphere lesion for one in the posterior fossa. In posterior fossa neoplasms the electroencephalogram usually shows diffuse slowing which may be maximal in both occipital regions (Figure 204). Although there may be diffuse slowing associated with some supratentorial tumors, in hemisphere lesions a more localized ab-

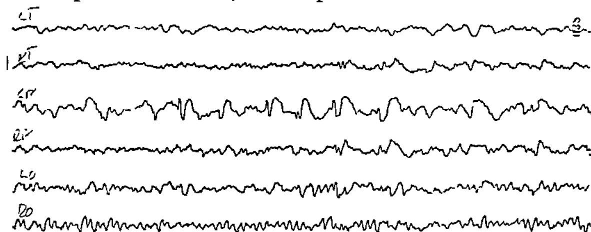


Figure 237. Electroencephalogram showing spike-and-wave focus in the left precentral area associated with a malignant glioma in the left fronto-parietal region of an 8 3/12 year old boy.

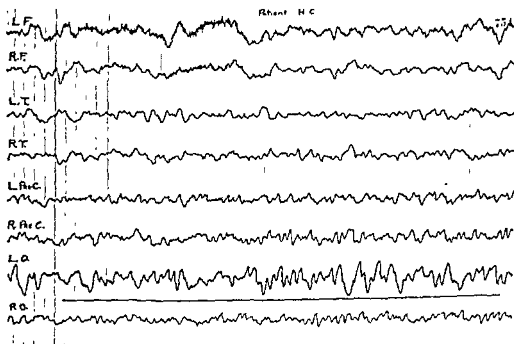


Figure 238 Eight year old boy with left occipital cystic astrocytoma. Note the well developed focus of three to four per second waves with increased amplitude in the left occipital lead Patient had had two convulsive seizures six months apart.

normality will almost invariably be present. In all but one of our cerebral glioma patients in whom electroencephalograms have been made the tracing has been grossly abnormal.

X-ray Examination

Roentgenographic studies of the skull should constitute an integral part of examination in every neurological diagnostic problem among infants and children. This is by far the most important diagnostic procedure avail-

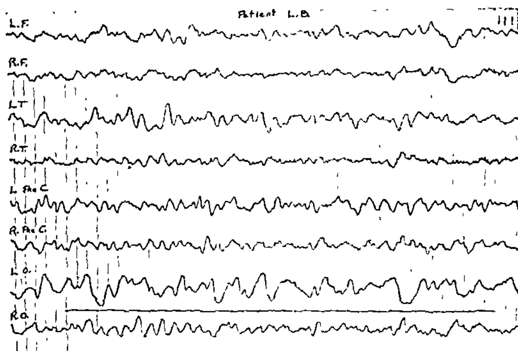


Figure 239. Electroencephalogram of three year old child with large partially cystic parieto-occipital ependymoma. There is a well developed focus of two to three per second waves with increased amplitude in the left occipital lead. The left temporal lead shows some indication of the same abnormality.

X-RAY FINDINGS IN 42 CEREBRAL GLIOMAS IN CHILDHOOD

Separation of Cranial Sutures	23
Normal Skull Films	12
Intracerebral Calcification	5
Positive Ventriculograms	24
Air in Cystic Tumors	10
Unsatisfactory Air Studies	4
Negative Air Studies	0

Figure 240. Positive findings on x-ray examination of children with gliomas of the cerebral hemisphere.



Figure 247A (*Upper*), and B (*Lower left*). Cystic astrocytoma of the right hemisphere in two and a half year old girl after aspiration of 60 ccs. of yellow fluid and injection of 2 ccs. of thorotrast and 5 ccs. of air. Note position of the solid tumor nodule posteriorly and inferiorly in the lateral view. Tumor subsequently totally removed.

C (*Lower right*). Lateral view postoperatively showing complete removal of solid tumor. Same patient as Figure 249





deep yellow in color. The protein content is high so that the fluid often gels quickly after removal. The tumor tissue is tough and relatively avascular. There is little necrosis and no calcification. There is no capsule but the tumor is often fairly well demarcated from the surrounding normal brain by its gross appearance and consistency (Figure 250).

Microscopically there is wide variation in the appearance of this tumor, but the usual picture is that of a homogeneous diffuse arrangement of astrocytes in a loose stroma without any definite pattern. The astrocytes have large oval or rounded clear nuclei and little or no discernible cytoplasm.



Figure 248. Carotid arteriogram in 9 5/12 year old boy showing marked displacement upwards and medially of the middle cerebral group of vessels. Patient had a large mixed glioma replacing most of the right temporal lobe. Satisfactory post-operative course after right temporal lobectomy.

A dense feltwork of branching cell processes is present. There are few mitoses and ordinarily little evidence of hemorrhage or necrosis (Figure 251).

Ependymoma: These tumors in the cerebral hemisphere arise presumably from the ependymal lining of the lateral ventricle and although the solid or cystic portions of the lesion may occupy any location within the hemisphere, at some point they encroach upon the ventricular lumen (Figure 252). They are firm, reddish-gray, often lobulated, and usually fairly well encapsulated. A high percentage of cerebral ependymomas in childhood are cystic. There may be multiple small cysts of varying size or a single cyst that may reach huge proportions (Figure 244). The cystic fluid is yellow to amber in color, often cloudy and often containing many red blood cells. These features usually distinguish it grossly from the clearer, lighter yellow fluid found in astrocytoma cysts. In ependymomas also, solid tumor usually lines the entire cyst wall projecting into the lumen in irregular nodules or shaggy, friable masses rather than remaining localized as a single mural nodule. Ependymoma has proved to be the commonest hemi-



Figure 249. Antero-posterior and lateral arteriograms in two and a half year old infant with a large cystic astrocytoma in the right internal capsular region. Note displacement upwards of the callosal branches of the anterior cerebral artery and displacement downwards and laterally of the branches of the middle cerebral artery. Same patient as shown in Figure 247.



Figure 250 Huge astrocytoma of the left cerebral hemisphere in 1 8/12 year old boy, partially cystic and partially solid.

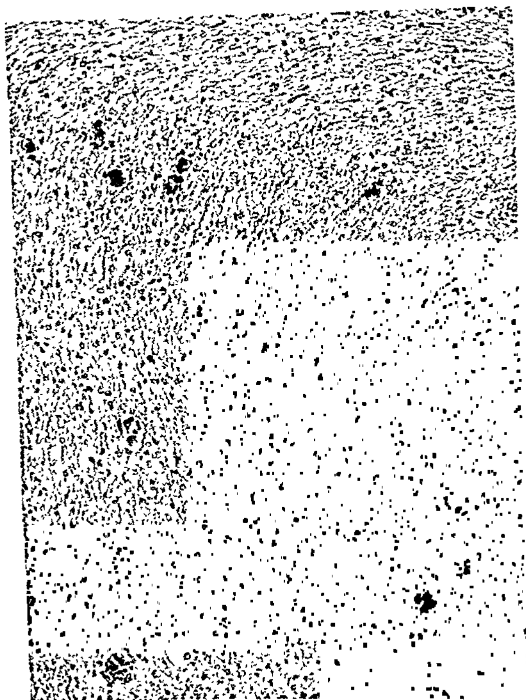


Figure 251. PTAH x 200 Photomicrograph of section from tumor nodule of cystic astrocytoma removed from the left occipital lobe of 7 11/12 year old boy. Note arrangement of the astrocytes in a loose fibrillary vascular stroma. There is no paravascular arrangement. The cells have large oval or round nuclei and poorly defined cytoplasm. The stroma contains numerous micro-cysts.



Figure 252. Huge ependymoma of the left cerebral hemisphere arising from the medial wall of the left lateral ventricle. Note enormous size to which this tumor has grown before death occurred.

sphere tumor to show calcification in our experience as well as that of others.[69]

Microscopically, these tumors are much less characteristic and uniform in appearance than ependymomas of the fourth ventricle. In the large lesions there are apt to be many areas of hemorrhage, necrosis and microcyst formation. The cells tend to be arranged in a paravascular distribution. The number of mitoses may vary considerably from one part of the tumor to another. The cells have fairly large oval nuclei, scant cytoplasm and tend to be quite uniform in size (Figure 253).

Glioblastoma Multiforme: Fortunately this highly malignant glioma is relatively much less frequent in the cerebral hemispheres in childhood than among adults. The tumor is rapidly growing, infiltrative and associated usually with late appearance of focal neurological abnormalities. Although these lesions may reach the surface by the time of surgery or post-mortem examination, they are deeply situated for the most part and they are never encapsulated (Figure 254). On cut section there is usually considerable degeneration, hemorrhage and formation of small cysts containing chocolate colored fluid. Some areas of the tumor may have the appearance of a much more slowly growing homogeneous glioma.

Microscopically the tumor is characterized by its heterogeneous ap-

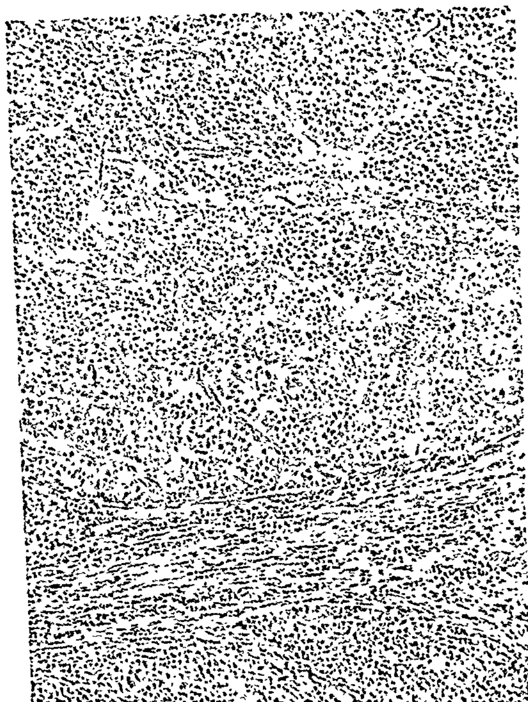


Figure 253 TBE x 200 Photomicrograph of section from recurrent ependymoma of the left parieto-occipital region in a 10½ year old boy. There are sheets of homogeneous cells with large oval nuclei and scant cytoplasm. Mitotic figures are common. *There is rarely any arrangement of cells with respect to blood vessels.* Fibrous tissue septa pass through the tumor. This tumor is much more like neuroblastoma now than in removals four years and two years previously.



Figure 254. [illegible] wing a huge glioblastoma [illegible] ving parietal

pearance and lack of organization. Cells vary greatly in size and arrangement. Multinucleated and atypical cells are frequent; mitoses are abundant. Areas of necrosis and hemorrhage are common. Blood vessels are numerous and show often a characteristic type of endothelial proliferation (Figure 255).

The pathological features of the less common cerebral hemisphere tumors in childhood will not be reviewed here in detail. Nor will any attempt be made to establish the criteria for separation of astroblastoma, ependymoblastoma, neuroblastoma, mixed glioma and cerebral medulloblastoma since it does not seem pertinent in a primarily surgical text. It may be of interest, however, to note one unusual tumor removed from the cerebral hemisphere of a one year old infant which seems to fit into the group of cases described by Russell, Marshall and Smith[171] as primary tumors of the microglia cells of the brain. Its resemblance microscopically to reticulum cell sarcoma as might be seen in a lymph node elsewhere in the body emphasizes the concept of the microglial cell as the reticulo-endothelial cell of the brain (Figure 256).

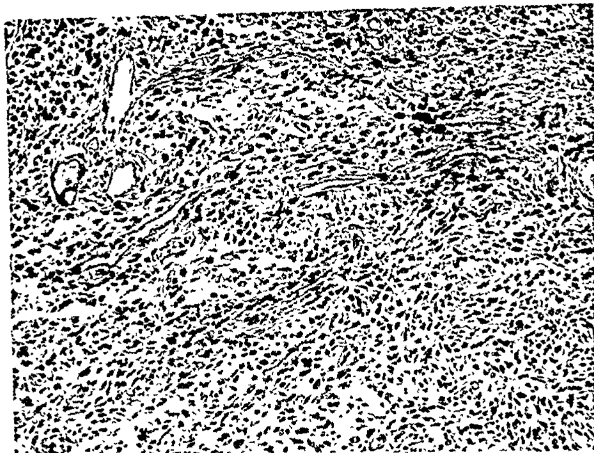


Figure 255. PTAH x 200 Photomicrograph from large glioblastoma multiforme largely replacing the right temporal lobe of a 9 4/12 year old boy. This is a very pleomorphic tumor with areas of hemorrhage, necrosis and minute calcification. In some places the tumor is loosely cellular and in others it is arranged in bundles and cords. The cells vary considerably in size and many abnormal shapes, multinucleated cells and mitoses are seen. There is a tendency to palisading around blood vessels.

Treatment and Results

As a whole, children with cerebral gliomas have been a discouraging group of patients to treat. Mention has already been made of the enormous size to which many of these tumors have grown by the time operation is carried out. Presumably this situation may be improved with more astute pediatric care and an earlier and wider application of newer objective diagnostic procedures.

At the present state of our knowledge the only significant treatment of these lesions is radical surgery. As with glioma surgery in adults, ultimate success is influenced by the histological type of the tumor, its location in silent or vital areas of the brain, and the adequacy and skill with which it is excised. Standard operative exposures and techniques are applied according to the localization determined pre-operatively.

In our series of cerebral gliomas all patients were operated upon except one infant who entered the hospital moribund and died within 48 hours

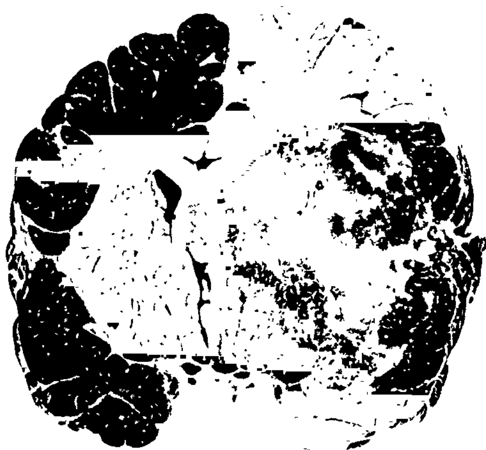


Figure 254. [illegible] wing a huge glioblastoma [illegible] ving parietal

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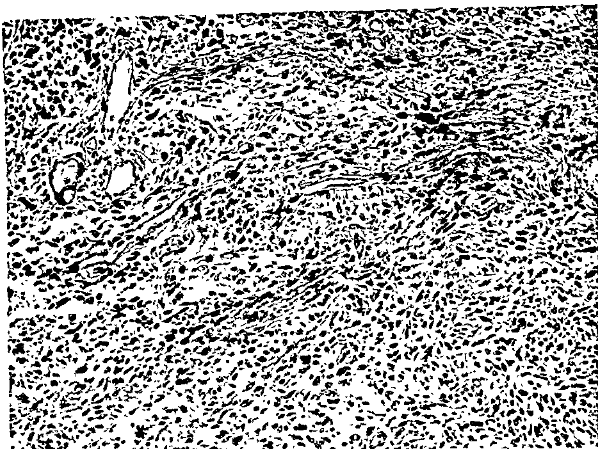


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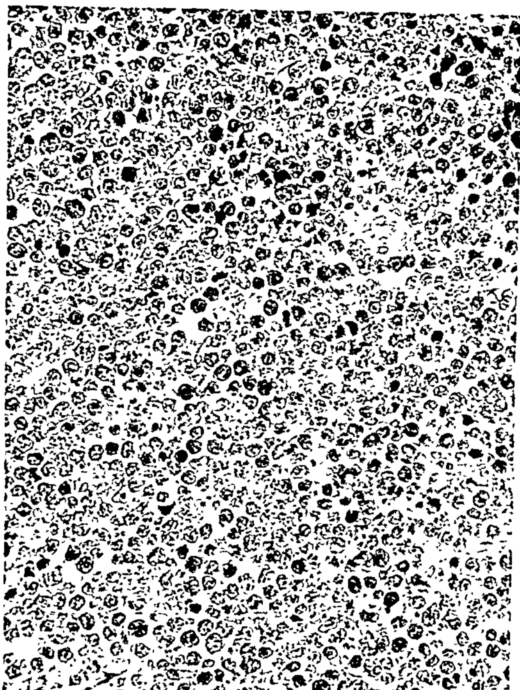


Figure 256. EMB × 400 Photomicrograph of section from huge nodular tumor in right fronto-parietal region of a one year old infant. The diagnosis is reticulum cell sarcoma (microgliomatosis) of the brain. The tumor consists of masses of uniform-appearing cells with large vesicular nuclei. These appear to be reticulum cells. Cells with pyknotic nuclei and mitotic figures are frequent.

after failing to improve on supportive therapy. In many instances, massive resections of these ordinarily enormous neoplasms were well tolerated (Figure 257). In a number of younger patients, the huge size of the lesion in itself made it seem unwise to attempt total excision at the time of primary operation. Re-operation seven to 14 days after the first resection was fre-



Figure 257. Large glioma (neuroblastoma) removed from the right fronto-parietal region of a 13 year old boy (400 grams of tumor removed). No evidence of recurrence up to 18 months.

quently rewarded by comparatively easy excision of further deeply placed tumor which had dislocated into the internal decompression of the original removal. With current methods of hemostasis, anesthesia and supportive therapy, massive one-stage resections in infants and small children are much more feasible. Attention to accurate replacement of blood lost and to regulation of body temperature throughout operation is particularly important.

In this series three patients died during the early post-operative period (two, five and 14 days); these should be considered deaths precipitated by the operative procedure. Seven patients died between two and six months after operation; these were for the most part patients in whom only a biopsy or minimal resection was carried out because of the nature of the tumor. Twelve patients died of recurrent neoplasm between seven and 30 months after initial treatment, and two patients from recurrent astrocytoma eight and 10 years respectively after the initial operation. Eighteen patients are still living from six months to 19 years after operation. Ten of the 15 patients in this series with astrocytomas are alive and as would be expected constitute the most favorable surgical results.

Anticonvulsant medication should be administered post-operatively to all patients with hemisphere tumors who have had seizures pre-operatively. This is continued usually for at least a year and indefinitely if clinical

seizures have recurred or the electroencephalogram shows seizure discharges.

In children with the more benign variety of gliomas (astrocytoma, ependymoma) incompletely removed, recurrent tumor should be treated by as radical surgical excision as possible, since prolonged remissions may frequently be obtained in this manner without undesirable additional loss of function.

Chapter IV

Craniopharyngioma

THE CRANIOPHARYNGIOMAS constitute one of the more common groups of supratentorial, but a relatively small proportion of the total intracranial tumors of childhood (Figure 194). The incidence of these lesions in reported series of neoplasms in children and early adolescents varies from five up to 13 per cent.[74, 101, 127] Among 313 intracranial tumors in children under 12 years of age, 21 craniopharyngiomas have been encountered in our clinic (Figure 258). It has been the commonest non-gliomatous intracranial tumor.

A number of different names have been applied to this tumor: suprasellar cyst, Rathke's pouch tumor, adamantinoma, cholesteatoma, hypophysial duct tumor and craniopharyngioma. The latter term, inaugurated by Cushing,[46] though not strictly accurate from an embryological point of view, is in widest general usage. Since the description by Erdheim in 1904[64] of the embryonic development of the hypophysis from union of two primordial components, one from the buccal epithelium (Rathke's pouch) and one from the primitive diencephalon, it has been known that squamous epithelial cell rests come to lie in the antero-superior region of the anterior lobe of the mature hypophysis. Presumably, proliferation of these squamous cell rests results in formation of solid masses of epithelial tissue and in cysts filled with keratin, cellular debris and cholesterol crystals. It is the progressive expansion of these solid and cystic lesions which results in compression of adjacent structures and produces such a grave symptomatology. The craniopharyngioma is a fascinating tumor because of its unique embryological origin and its diverse clinical features; it is a challenging tumor to the neurosurgeon because of its encapsulation, frequent cystic nature and susceptibility to accurate pre-operative diagnosis and localization; at the same time, it is a baffling and discouraging lesion because its location defies complete removal without undue damage to vital surrounding structures.

Although craniopharyngiomas may become symptomatic at any period of life, they are most commonly discovered during the first two decades. The clinical picture varies considerably in different age groups. In childhood the most prominent symptoms and signs are apt to be related to increased intracranial pressure and growth impairment, in young adult

life to abnormalities of endocrine and metabolic function, and in older patients to visual disorders. However, all of these features may be present in any one patient at any age. Here, we are concerned principally with its *diagnosis and management in early life.*

Symptoms and Signs

The suprasellar location of this tumor makes possible a number of different physiological disturbances. These can be divided into four groups *according to the adjacent structures which are involved:* (1) increased intracranial pressure; (2) visual disorders; (3) pituitary and other secondary endocrine dysfunction; and (4) disturbances of hypothalamic function.

Increased Intracranial Pressure: In young children especially, headache and vomiting frequently constitute the initial complaints. These are a result of internal hydrocephalus and may differ very little from the same complaints seen so frequently in mid-line posterior fossa neoplasms. The headache is usually generalized and accompanied by irritability. Vomiting may be forceful and sudden and is particularly prone to occur in the early morning. Papilledema may be present, as well as a "cracked-pot" percussion note to the skull, sixth nerve weakness, dilatation of the scalp veins and other manifestations of increased pressure. Internal hydrocephalus is limited to one or both lateral ventricles and is due to obstruction of the

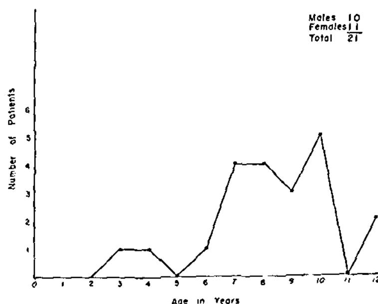


Figure 258 Age and sex distribution of children with craniopharyngiomas Only two children under six years of age have been treated

anterior third ventricle or one or both foramina of Monro by upward expansion of the craniopharyngioma. Eighteen of the 21 children in this series showed evidence of increased intracranial pressure.

Visual Disorders: The optic chiasm, optic nerves, or optic tracts may be compressed from a variety of angles producing diminution of visual acuity in one or both eyes and producing many different types of visual field defects. *Since a mid-line suprasellar location of the tumor is common, it is* natural to find visual defects frequently in the temporal fields. This is usually

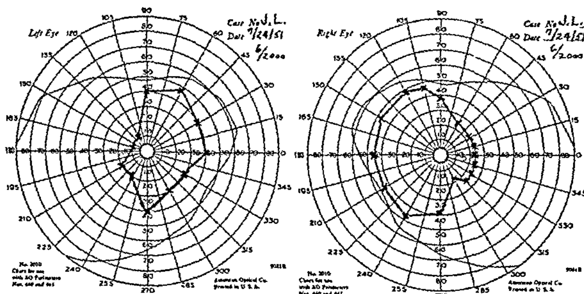


Figure 259. Visual fields in four years and seven months old girl with craniopharyngioma. Diagnosis had been made on incidental skull films made because of a head injury. This bitemporal visual field defect appeared suddenly at a routine three months check-up. Patient otherwise asymptomatic.

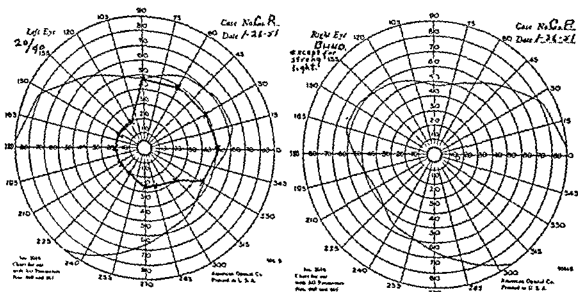


Figure 260. Visual fields in an eight year old girl with a huge, partially solid, partially cystic craniopharyngioma. Vision in the right eye was too poor to measure. There is a temporal field defect in the left eye and diminished acuity in the remaining nasal field of vision.

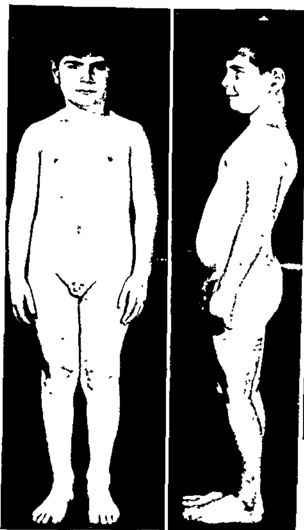


Figure 261. Characteristic habitus of 7 2/12 year old boy with craniopharyngioma. Note small size of external genitalia.



Figure 262. Eight year old girl with craniopharyngioma showing marked retardation of growth but no impairment of mental development. On the right is a normal girl of the same age.

partial and asymmetrical in early stages (Figure 259), and later characterized by bitemporal. hemianopsia. If one optic nerve is primarily involved, scotomata are present and complete blindness of that eye may ensue (Figure 260). If one optic tract is damaged, homonymous hemianopsia results. When the optic pathways are compressed and there is little or no increased intracranial pressure, opthahalmoscopic examination reveals primary optic atrophy. Visible optic atrophy is usually accompanied by marked loss of visual acuity. Obviously, in young children minor impairment of visual acuity and the presence of small field defects are virtually impossible to determine. Fourteen of the 21 patients in this series demonstrated visual disturbances at the time of hospitalization.

Pituitary Dysfunction: Suppression of normal pituitary activity in childhood is manifested principally by retardation of growth. This is noted



Figure 263. Eight year old boy with craniopharyngioma. Note small stature as compared with normal boy of same age on the left. Patient entered the hospital because of failing vision and failure to grow. Mental development is normal for his age.



Figure 264. Five years and eight months old child with advanced hypopituitary syndrome (Simmond's disease) secondary to large craniopharyngioma.

by smallness of stature, arrested skeletal and tooth development or sexual infantilism (Figure 261). These children may be well proportioned but dwarfed beside a normal child of the same chronological age (Figures 262 and 263). Since they may remain mentally quite alert, it is sometimes startling to find the ideas and reactions of a 10 to 12 year old coming from a child who appears to be only six or seven by his size. Pale, smooth, hairless, fine-textured skin is usually present. An early complaint is increased lassitude and easy fatiguability. These children lose the inexhaustible supply of energy characteristic of the latter half of the first decade; their reserves are meagre and difficult to mobilize. The basal metabolic rate is often considerably below normal. Blood pressure may also be low. In

advanced lesions, the picture of pan-hypopituitarism (Simmonds' cachexia) may become extreme (Figure 264). Fifteen of the 21 children in this series showed evidence of pituitary dysfunction.

Hypothalamic Disorders: If the tumor extends upward to invade the floor of the third ventricle, structures of the hypothalamus in its walls may be compressed. Drowsiness in the absence of increased intracranial pressure is indicative of posterior hypothalamic involvement, as is the presence of persistently subnormal temperature. Polyuria and polydipsia (diabetes insipidus) are indicative of compression of the supra-optic and paraventricular nuclei of the anterior hypothalamus, or of the tract connecting these nuclei with the posterior lobe of the pituitary. This is occasionally seen as an early complaint in craniopharyngioma (three patients) and frequently as a



Figure 265. Lateral roentgenogram of the skull in a six year old girl with craniopharyngioma Note separation of the coronal and lambdoid sutures and mottled area of calcification just above the slightly enlarged sella turcica (Reprinted through the courtesy of The C V. Mosby Company from *J. Pediat*, 29.95-116, 1946)

transient disorder following operation. *Adiposo-genital dystrophy is uncommon in children with craniopharyngioma, but when present indicates* tuber cinereum involvement. These youngsters with hypothalamic encroachment may sometimes experience periodic attacks characterized by semi-stupor, fever, cardiac irregularities, low blood pressure and constriction of the pupils.

X-ray Findings

Roentgenograms of the skull are abnormal in almost every child with a craniopharyngioma, and they are conclusively diagnostic in a high percentage of cases. *Delicate, diffuse flecks of calcium in the intra- or suprasellar region of a child,* particularly in the presence of signs of increased intracranial pressure or enlargement of the sella, are pathognomonic (Figure 265). Such calcification is discernible in 60 to 70 per cent of patients in most series.[141] The calcification *is usually in the solid portion of the lesion* and does not indicate what the *size of a cyst* or the whole *tumor*



Figure 266. Lateral roentgenogram of the skull in eight year old boy after aspiration of craniopharyngioma cyst through the coronal suture and injection of air into the cyst. Note that the cyst is above and behind the calcified area which is presumably in solid tumor The sella turcica is slightly enlarged.

may be (Figure 266). Sixteen of the 21 children in this series showed suprasellar calcification.

In approximately 50 per cent of patients, the sella turcica itself is enlarged or distorted (Figure 267). It is wise to remember, however, that a craniopharyngioma may be present, especially in a young child, with an entirely normal appearing sella and with no visible calcification on x-ray examination. In this series of 21 children, 12 showed an abnormality of the sella turcica and in nine it appeared normal by x-ray examination. Roentgen evidence of increased intracranial pressure may be present: separation of the cranial sutures, erosion of the posterior clinoid processes and increase in convolutional markings (Figure 265).



Figure 267. Lateral roentgenogram of the skull in 11 year old girl whose bone age was that of a child of six and who showed a bitemporal hemianopsia. Note enormous size of the sella turcica due to cystic craniopharyngioma.

Ventricular air studies are helpful in diagnosis, in determining the size and upward extent of the lesion and in estimating the degree of internal hydrocephalus as well as the presence or absence of an interventricular block. If a large cyst protrudes into the anterior third ventricle, its margins can usually be demonstrated clearly (Figure 268A). Ventriculographic demonstration, though usually unnecessary to make the diagnosis, is frequently of great value in planning surgical treatment. Air may also be introduced directly into the cyst after tapping through a burr hole (Figures 266 and 268B).

Laboratory Findings

Subnormal fasting blood sugar, flattened glucose tolerance curve, and low basal metabolic rate may all be seen. Ordinary blood and urine examinations are usually normal. High total circulating eosinophil counts and poor falls in this count in response to adrenalin and ACTH test doses are indicative of impaired pituitary and adrenal functions.[194] Depressed 17-ketosteroid excretion also indicates poor adrenal function, which in these patients is indicative of depressed pituitary stimulation.

Morbid Anatomy

There is tremendous variation in the size, extent, and gross appearance of craniopharyngiomas. They are almost invariably cystic to some degree.

The cyst may be tiny and surrounded by masses of solid tumor; it may be multiloculated and irregular in shape; it may be a huge single cyst projecting upward into the third ventricle, laterally into the frontal or temporal lobe or posteriorly into the interpeduncular area (Figure 269). The solid portion of the tumor in any case is apt to be inferior to the cyst and closely applied to the antero-superior aspect of the pituitary gland (Figure 270). The tumor may be located mostly within an enlarged sella turcica or it may be entirely above the diaphragma sella.

The capsule of the cyst is usually smooth, grayish-white externally and sometimes very tough in consistency. This capsule is derived from the tumor's connective tissue stroma. It adheres closely often to surrounding structures – hypothalamus, dura, optic chiasm, arteries of the Circle of Willis, pituitary gland – making surgical dissection extremely difficult. The cyst contains desquamated and liquefied epithelial debris. This fluid varies in color from light yellow to dark golden-brown and has a characteristic oily appearance. Suspended in the fluid are numerous cholesterol crystals which readily reflect transmitted light.

The solid portion of the lesion may be necrotic or may be very tough and vascular. Calcification is present in the solid tumor grossly in a high percentage of cases; less frequently, a thin layer of calcification occurs in the cyst capsule.

Microscopically, the great majority of our cases in young children fall into Bailey's[9] classification of simple squamous epitheliomas. The tumor



Figure 268A (*Left*). Lateral roentgenogram after ventricular injection of air demonstrating hydrocephalus with absence of the right frontal horn and displacement of the anterior third ventricle.

B (*Right*). Same patient after aspiration of enormous craniopharyngioma cyst (165 ccs. of oily fluid) and injection of air into the cyst lumen. Suprasellar calcification is also visible as shown by arrow. Same patient as shown in Figure 264.

consists of irregular masses and strands of stratified squamous epithelium in a stroma of fibrous connective tissue (Figure 271). There is keratinization of the layers of cells nearest the cyst lumen. Minute areas of calcification are frequent. More rarely, adamantinomatous types of craniopharyngioma occur in which palisades of columnar cells resembling the adamantoblasts of tooth buds are found adjacent to connective tissue. Mitosis, blood vessel invasion, and infiltration of surrounding structures by tumor cells are not seen.



Figure 269. Post-mortem view of the base of the brain showing large craniopharyngioma. Solid portion of the tumor has grossly distorted the optic chiasm and optic tracts. A large cyst which is collapsed in this picture extended caudally through the incisura into the posterior fossa. Patient was blind, had communicating hydrocephalus and developed tonic seizures and decerebrate posture before death.

Treatment

Total surgical removal is obviously the ideal treatment. However, due to the tumor's close anatomical relationship with the pituitary gland and the hypothalamus this is rarely possible. Particularly in young children, attempts at complete excision have carried a very high mortality rate because of acute hypopituitary and hypoadrenal crises. Although it may be possible to dissect out the tumor completely without directly injuring the pituitary gland or the hypothalamus, their blood supply is evidently compromised under such circumstances in a high percentage of cases. Since there is no known method of replacing the growth hormone of the pituitary gland, total removal of the gland in childhood is undesirable. It is our feeling, therefore, that although total excision can undoubtedly be accomplished in rare instances, its attempt is not ordinarily warranted.

Since the tumor produces symptoms by compression of neighboring structures, treatment other than total excision is directed toward reduction of the mass of the lesion and re-establishing spinal fluid circulation.

If the patient's symptoms are primarily those of pituitary suppression, if the sella turcica is enlarged, and there is no evidence clinically or by



Figure 270 Post-mortem longitudinal section of the brain of 10 year old girl with craniopharyngioma. Note solid portion of the tumor just above the compressed pituitary gland and the huge cyst extending forwards into the frontal lobe and upwards into the anterior third ventricle. (Reprinted through the courtesy of The C. V. Mosby Company from *J Pediat.*, 29:95-116, 1946.)



Figure 271 EMB x 200 Photomicrograph from solid portion of craniopharyngioma surgically removed from third ventricle of an eight year old boy. Note the characteristic loosely arranged edematous fibrous connective tissue containing masses of stratified squamous epithelial cell rests. There is considerable calcification. Note the keratin pearls lining the surface of a cyst membrane.

ventriculography of obstruction to the anterior third ventricle, it is probable that the tumor is largely solid and is largely intrasellar in location. Such a tumor is exposed directly by craniotomy. It is our custom to use a coronal incision which permits a very low-placed frontal bone flap to be elevated (Figure 305). This is done on the side which has shown the greatest visual impairment or on the right side if there is no marked visual asymmetry. The frontal lobe is elevated as spinal fluid is removed through a lumbar puncture needle until the optic nerve and prechiasmatic region are exposed. Every attempt is made to avoid compression of or traction on the suprachiasmatic area. If a cyst is identified it is aspirated and the capsule opened after carefully teasing it away from the optic nerves and any adherent vessels. As much of the capsule above the sella turcica is removed as possible together with any solid tumor that is easily mobilized. No attempt is made ordinarily to remove the inferior portion of the capsule if it is within the sella turcica or to remove any tissue beneath or posterior to the capsule of the cyst.

If the patient has internal hydrocephalus and a mass is demonstrated projecting upward and posteriorly into the third ventricle by ventriculography, it is assumed that this mass is a large cyst. It is our custom then to introduce a needle through the coronal suture or through a drill hole into the cyst and evacuate its contents. If such a cyst cannot be aspirated this way under x-ray guidance it may be necessary to do a craniotomy and approach the cyst subfrontally or transventricularly. If drainage of a cyst projecting into the third ventricle does not relieve the symptoms and signs of increased intracranial pressure, a unilateral or bilateral Torkildsen procedure (p. 136) is performed to side-track the block of spinal fluid circulation (Figure 272). Repeated "blind" aspiration through a drill hole may be carried out at intervals if the symptoms recur. If simple aspiration of a cyst does not relieve impairment of visual acuity or visual fields, then direct subfrontal exposure should be performed. Conservative treatment of this kind may give relief of increased pressure and preservation of vision for many years.

If pre-operative studies show marked suppression of pituitary function it may be expected that the post-operative adrenal response will be poor no matter whether the pituitary gland is further compromised by the operation itself or not. This may be treated by protecting the patient through the operation and post-operative period with cortisone, or by stimulating the patient's own adrenals to more normal function by a course of ACTH prior to surgery.[89] In this clinic eight consecutive children with craniopharyngiomas treated in this manner have had unusually smooth, uncomplicated operative and post-operative courses.

Cortisone has the advantage of quick action. It should be started no



Figure 272. Antero-posterior and lateral roentgenograms after bilateral Torkildsen procedure as performed for craniopharyngioma blocking the anterior third ventricle.

more than 24 hours before surgery and discontinued within a week in order to minimize interference with wound healing. A tentative schedule for the administration of cortisone acetate is suggested as follows:

100 mgm. on night before operation
100 mgm. on morning of operation
25 mgm. every 6 hours for 1 day post-operatively
25 mgm. every 8 hours for next 2 days
25 mgm. every 12 hours for next 2 days
25 mgm. per day for next 2 days.

The intramuscular route should be used because of the more prolonged effect as compared with oral administration.

If ACTH is employed, more prolonged pre-operative administration is required. It should be started at least five days before surgery to obtain adequate activation of the partially involuted adrenal cortex and insure sufficient circulation of adrenal hormones at the time of maximal stress. Here, too, dosage should be reduced as soon as possible after surgery. The following schedule is suggested for administration of this drug:

20 mgm. every 6 hours for at least 5 days pre-operatively
20 mgm. every 6 hours for 2 days post-operatively
20 mgm. every 8 hours for next 2 days
20 mgm. every 12 hours for next 2 days
20 mgm. during next 24 hours.

Patients who show continued suppression of pituitary function may

need to be carried on small maintenance doses of cortisone (25 mgm. daily), added salt, and occasional small doses of desoxycorticosterone (1 to 2 mgm.) indefinitely.

Although from a histological point of view there is no particular reason to think that deep x-ray therapy should help this lesion, it has been thought by some that a definite response has been seen. It seems unlikely that x-radiation would have any effect on the growth of stratified squamous epithelium, but perhaps the rate of cystic fluid formation has been altered.

Prognosis

The results in treatment of craniopharyngiomas in young children are not as good as when the lesion makes itself known first in the older age groups. This is probably true because of the frequency of third ventricular extension with internal hydrocephalus and hypothalamic encroachment in the pediatric age period. Eight of the 21 patients in this series are living — from six months to 10 years after initial surgery. With the availability of cortisone and ACTH, crises of the operative and post-operative periods should be minimal, and normal adrenal function subsequent to pituitary damage easier to maintain. Although complete cure will undoubtedly always be rare, careful surgery should result in a high percentage of these patients in prolonged remission of visual impairment and increased intracranial pressure. Evidence of pituitary regeneration following surgical treatment has been infrequent.

Chapter V

Tumors of the Choroid Plexus

Papilloma and papillary adenocarcinoma of the choroid plexus constitute one of the more unusual and interesting types of intracranial neoplasm seen in infants and children.[18, 63, 86, 165] In our clinic, nine children varying between the age of two weeks and 21 months have been treated (Table X).

Clinical Features

The tumor arises in one of two locations; either the glomus of the choroid plexus in the lateral cerebral ventricle or the choroid plexus of the anterior medullary velum of the fourth ventricle. To our knowledge none has been reported in the anterior portion of the lateral ventricles or in the third ventricle. Since these tumors do not invade neural tissue, they do not give rise ordinarily to localizing symptoms and signs. Their presence is made known either by increased intracranial pressure or occasionally by hemorrhage into the circulating cerebrospinal fluid.

Choroid plexus papillomas of the lateral cerebral ventricle are unusual because they are associated with a communicating type of hydrocephalus;[106] that is, there is diffuse enlargement of the subarachnoid pathways including both lateral ventricles, the third and fourth ventricles and the basilar cisternae. It seems probable that they are accompanied by increased formation of cerebrospinal fluid since there is no evidence of obstruction along the spinal fluid pathways and no evidence of impaired absorption. Following simple removal of the tumor, signs of increased intracranial pressure disappear. It is perhaps possible that diffuse enlargement of the ventricular system is due simply to the effect of the large pulsating tumor mass itself.

Choroid plexus tumors within the fourth ventricle may give rise to internal hydrocephalus by increased formation of cerebrospinal fluid, but in addition tumors in this location also cause direct obstruction of the fourth ventricle or aqueduct. This cannot usually be differentiated previous to operation from other types of obstructive internal hydrocephalus.

X-ray Examination

The diagnosis of choroid plexus papilloma of the lateral ventricle is usually made by ventriculography. Plain roentgenograms show only evi-

Table X

CHOROID PLEXUS TUMORS

No.	*Name*	*Age*	*Presenting Symptom*	*Location of Tumor*	*Operation*	*Pathological Diagnosis*	*Result*
1.	C. T	15 days	Enlargement of head	Rt. lateral ventricle	Total removal	Papilloma	8 months Asymptomatic
2.	C. D.	4 months	Bulging fontanelle	Rt. lateral ventricle	Total removal	Papilloma	14 months Asymptomatic, slightly retarded
3.	P. S.	14 months	Vomiting	Fourth ventricle	Total removal	Adenocarcinoma	5 days post-op. Died, rupture of stomach
4.	K. L.	9 months	Vomiting	Lt. lateral ventricle	Incomplete removal	Adenocarcinoma	3 years and 1 month Asymptomatic
5.	R. S.	8 months	Convulsion	Rt. lateral ventricle	Total removal	Papilloma	11 years Asymptomatic
6.	D. D.	21 months	Unsteady gait	Rt. lateral ventricle	Incomplete removal	Adenocarcinoma	5 months post-op. Died
7.	N. L.	4 months	Enlargement of head	Lt. lateral ventricle	Incomplete removal	Adenocarcinoma	2 weeks post-op. Died
8.	L. B.	9 months	Vomiting and enlargement of head	Lt. lateral ventricle	None	Papilloma	Died unoperated (post-mortem finding)
9.	S. M.	19 months	Unsteady gait	Lt. lateral ventricle	Incomplete removal	Adenocarcinoma	Moribund at time of operation. Died during attempted excision

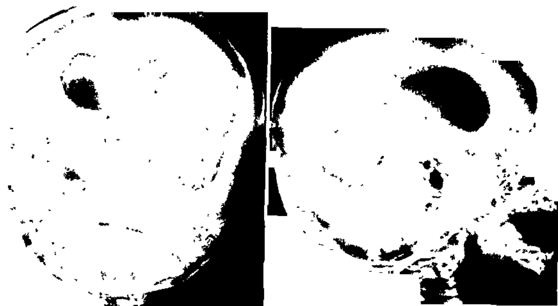


Figure 273. Postero-anterior and lateral ventriculograms of six months old infant showing large papilloma of the choroid plexus. Note dilatation of the entire ventricular system.



Figure 274 Lateral ventriculogram showing marked symmetrical dilatation of the lateral and third ventricles. Note telescoping of the aqueduct and dilatation of the fourth ventricle as well *The irregular defects in the posterior wall of the fourth* ventricle represent indentations due to the tumor itself, papillary adenocarcinoma of the choroid plexus.

dence of enlargement of the head in infants or of increased intracranial pressure in older children. These tumors do not calcify. Ventricular air studies, however, may be extremely dramatic due to demonstration of the intraventricular mass standing out in sharp contrast to the air *surrounding* it (Figure 273). Ventriculograms also demonstrate symmetrical diffuse dilatation of the entire ventricular system in spite of the limitation of the tumor to the atrial region of one lateral ventricle.

In choroid plexus tumors of the fourth ventricle, the ventriculographic picture is that of obstructive internal hydrocephalus. With good filling and particularly fortunate films, it may be possible to demonstrate the actual tumor mass within the lumen of the fourth ventricle (Figure 274).



Figure 275. Papilloma of the choroid plexus of the lateral ventricle removed successfully from six months old infant. Post-operative course uneventful to date (one year).

Morbid Anatomy

This tumor may grow to large proportions within the lumen of the lateral ventricle, even in small infants (Figure 275). Grossly, the tumor is irregular, reddish-purple in color, soft, but well contained within a thin transparent capsule. It does not invade neural tissue, nor adhere to the ependyma of adjacent ventricular walls. It shrinks in size considerably when the blood supply has been secured. In the fourth ventricle it is more the shape of the ventricular lumen and may extend out through the lateral recesses into the basilar cisternae.

Microscopically, the cell pattern of these tumors is that of the choroid plexus itself (Figure 276). In some instances there is evidence of blood vessel invasion and increased mitotic activity near the tumor's attachment. Under these circumstances the term papillary adenocarcinoma of the choroid plexus is perhaps more appropriate. Evidence of infiltration beyond the mass in the ventricle or of distant metastases has not, however, been seen.

Treatment and Results

Treatment consists of complete surgical excision wherever possible. This can usually be accomplished successfully when the lesion occurs within the lateral ventricle. Since the tumor is located in the atrial portion of the ventricle, it is approached in a manner similar to that for per-

forming choroid plexectomy (Figure 130). A straight or horseshoe shaped scalp incision is made so that a small bone flap can be elevated in the posterior temporo-occipital region. The ventricle is exposed by a transcortical incision through an avascular area. Malleable retractors introduced into the ventricular lumen support the brain and maintain exposure during re-



[illegible] papillary adenoma [illegible] fairly regular pseudo-stratified columnar epithelium. The nuclei are basally placed and supranuclear vacuoles are prominent. There is no evidence of malignant change. Same tumor as Figure 275.

moval of the tumor (Figure 277). The tumor is usually attached in the region of the glomus of the choroid plexus. This attachment can sometimes be secured and bleeding avoided with silver clips and cautery so that the tumor is removed intact (Figure 275). The operation is usually well tolerated and not accompanied by post-operative complications if intraventricular hemorrhage is controlled.

In the fourth ventricle, complete excision is more difficult because the exposure is more hazardous and because the tumor tends to extend through the lateral recesses of the fourth ventricle to inaccessible areas anterior to the brain stem. If the tumor is limited to the lumen of the fourth ventricle, however, complete removal should be possible.

If microscopic studies suggest adenocarcinoma in any part of the tumor or if complete removal has proved impossible for any reason, deep x-ray therapy to the tumor site is recommended. A tumor dose of 2,000 to 3,000 R has been used.

In our experience the outcome has been quite variable. Survivals of eight months to 11 years without recurrence have occurred in four patients. Certainly every effort should always be made to perform complete excision as soon as the diagnosis is made.



Figure 277. Operative photograph showing the brain being held up with malleable retractors during intraventricular exposure for removal of a choroid plexus tumor.

Chapter VI

Tumors of the Skull and Meninges

CONGENITAL TUMORS

Clinical Features

IT WOULD PERHAPS be more accurate to describe the common dermoid and epidermoid cysts as tumors of the scalp. However, they come into the field of the neurological surgeon because frequently in children these congenital inclusion cysts encroach upon or lie within the skull. They are always connected to the over-lying skin and *in almost every instance if the* shaved skin is examined carefully in good light a tiny dimple or point of skin fixation can be identified.



Figure 278 Typical appearance of dermoid inclusion cyst in the anterior temporal region In this location these cysts very often penetrate through the skull but they do not go through the meninges

There is great predilection in *infants and children for congenital* inclusion cysts to occur in the supra-orbital and anterior temporal regions (Figure 278). They occasionally occur elsewhere over the vertex (Figure 279). When a cyst occurs in the mid-line either at the *bridge of the nose or in the occipital* region, it is of particular importance because of the increased incidence in these locations of intracranial extension, as discussed elsewhere (p. 71).

The scalp swelling produced *by the cyst is rubbery hard, non*-tender, and sometimes partially movable. If the cyst is imbedded in the skull, however, it may be



Figure 279 One year and eight months old boy with epidermoid cyst in the region of the anterior fontanelle. This cyst extended through both layers of the cranium but did not penetrate the dura.

fixed to palpation. The cyst may become infected, in which case redness, edema and tenderness are present and there may be a thin or frankly purulent discharge onto the scalp surface. Osteomyelitis may occur (Figure 334).

The x-ray appearance of those cysts which extend into the skull is characteristic (Figure 280). A smooth rounded area of decreased density is seen, the margin of which is regular and usually shows a thin line of increased density. In a tangential view it may be possible to determine whether the cyst has eroded only the outer table of the skull, into the diploë, or through the inner table as well.

The cyst consists of a fibrous capsule of variable thickness which contains cellular debris, and often sebaceous material and hair. The cyst is lined by stratified squamous epithelium which frequently on histological study shows evidence of chronic inflammation.

Treatment

Dermoid and epidermoid cysts involving the skull should be treated promptly by complete surgical excision, both for cosmetic reasons and as prophylaxis against infection either in the scalp, the bone, or intracranially. Excision before infection has occurred is simple and healing is prompt. If



Figure 280. Lateral roentgenogram of the skull of a two year old infant showing area of diminished density surrounded by a sharp border of increased density. This is characteristic of an epidermoid cyst protruding through the outer table of the skull.

x-ray examination has revealed a bony defect, particularly in the mid-line (Figure 64), operation should be planned to trace and remove an intracranial extension if this should be found.

Ordinarily a small straight or elliptical incision is made directly over the mass. If a skin dimple is present, it should be excised. Incision is carried down to the capsule and dissection continued around it to remove the mass if possible without spillage of its contents. The entire bed of the cyst should be inspected carefully to insure the absence of any stalk extending intracranially. Frequently the cyst rests on the dura, or there may be a thin shell of inner table of the skull remaining.

After complete removal of the cyst, if the bony defect is a centimeter or more in diameter, it is wise to raise some "curls" of bone from the adjacent outer table and reflect them into the lumen; solid bony healing will then ordinarily ensue. A larger defect may occasionally need some more elaborate type of cranioplasty (p. 173).

More complex tridermal congenital tumors involving primarily the scalp and skull are also occasionally seen (Figure 281). The earliest possible total



Figure 281A (*Left*). Huge partially cystic and partially solid teratoma involving the scalp, face, orbit and middle cranial fossa in a new-born child. No evidence of malignancy. Normal pneumoencephalogram

B (*Right*). Four months after total excision. Patient developing well. Negative neurological examination except for paralysis of the musculature of the left upper lip and cheek.



Figure 282. Teratoma of the side of the cranial vault and face completely removed at operation. The tumor has been opened showing the variegated appearance of the interior. The solid portion of the tumor contained bone, cartilage, muscle, epithelium and fibrous tissue. Same patient shown in Figure 281.

excision of such lesions is recommended, not only for cosmetic reasons but because of the tendency toward malignant change in these abnormal tissues (Figure 282).

EOSINOPHILIC GRANULOMA

Clinical Features

The etiology of this interesting and rare granulomatous lesion which affects the bones primarily of children and young adults is still unknown. Jaffé and Lichtenstein [103, 122] feel that it is a peculiar type of inflammatory histiocytosis which is a milder form of the same generalized disease process seen in Hand-Schüller-Christian and in Letterer-Siwe disease. An infectious agent has never been demonstrated. Since the lesions are often single and associated with a history of injury, the etiological significance of trauma has been suggested but never proved.[151]



Figure 283 One year old child with eosinophilic granuloma of the right temporal bone. Overlying soft tissue mass had been mistakenly incised as an abscess before hospital admission. Chronic sinus resistant to healing remains.

In a high percentage of all patients with this disease the bones of the cranial vault are involved by one or more lesions.[76] This may be the only location or there may be simultaeous foci in the long bones. In any case, demonstration of a single lesion should always be followed by survey x-ray examination of the entire skeleton. This skeletal survey should be repeated at intervals of six months for several years, even if clinical examination remains negative.

Since there is rarely evidence of fever, malaise or other systemic disturbance, the lesions are usually disclosed because of local symptoms and signs, or else they are uncovered incidentally on x-ray films made for some other purpose. A slight palpable swelling is usually present over lesions in the cranial vault. This area is tender and there may be edema of the surrounding scalp. The swelling is soft but not fluctuant. There may be local heat so that an abscess of the scalp or osteomyelitis is suspected (Figure 283). If the lesion involves the supra-

orbital ridge, there may be ptosis and slight exophthalmos. Although low-grade eosinophilia in the peripheral blood stream has been reported, examination of the circulating blood ordinarily reveals no abnormalities.

The x-ray appearance is characteristic but not in itself diagnostic. Skull lesions usually appear as roughly circular, punched-out areas of decreased density. They may be found in any of the bones of the cranial vault and may vary from a few millimeters to several centimeters in diameter. The lesion may become considerably larger or smaller within a week or two, even in the absence of any treatment. The edges of the bony defect are not as smooth as in defects due to dermoid cysts or other congenital lesions and there is not the narrow line of increased density at the margin (Figure 284). By x-ray examination alone, eosinophilic granuloma cannot be distinguished from localized osteomyelitis, myeloma, sarcoma or Hand-Schüller-Christian disease.



Figure 284. Lateral roentgenogram of the skull of an eight year old girl showing characteristic punched-out lesion of the parietal bone due to eosinophilic granuloma. Proved by biopsy and treated successfully with local x-radiation.

Morbid Anatomy

The gross features of this tumor suggest that it is either a very vascular type of chronic inflammatory granulation tissue or a necrotic, cellular neoplasm. It is soft and suckable and easily curetted from adjacent bone. It erodes through both cortices of the skull but does not penetrate the dura. Cultures made at the time of surgical treatment are sterile.

Microscopically this is a highly cellular, vascular lesion, often with large areas of necrosis and very little stroma (Figure 285). There are many large mononuclear phagocytic cells, but the characteristic feature is the overwhelming abundance of eosinophils. Bacteria are not seen. Adjacent bone shows evidence of both destruction and healing.

Treatment and Results

The diagnosis of eosinophilic granuloma can be made only by biopsy. In young children no further surgical treatment is ordinarily necessary, since once the diagnosis is confirmed, local deep x-ray therapy causes rapid resolution of the lesion and bone healing promptly ensues. As a matter of practice, biopsy usually consists of suction and curettage of most or all of the soft tissue granuloma. Block resection of an area of the skull is unnecessary. If subsequent lesions appear elsewhere in the skull they may be treated directly by x-ray therapy.

X-ray therapy is administered in small doses to each local lesion. Six hundred to 900 R (tumor dose) given over a three to four day period is usually sufficient to bring about satisfactory disappearance of the lesion. The prognosis for ultimate complete remission is good, particularly when the disease is limited to the skull. Occasional lesions heal spontaneously, but in general healing is achieved more promptly and effectively by these small doses of local radiation.

FIBROUS DYSPLASIA

Clinical Features

Fibrous dysplasia of bone is a disease of unknown etiology which is commonest in childhood and characteristically becomes less active or arrested as the period of active skeletal growth is completed. It is probably not a metabolic or endocrine disorder as often suggested but a developmental mesenchymal defect characterized by accumulation of masses of fibrous tissue in one or more bones.[123] There is no proof that it represents a peculiar type of reparative response to local injury. When there are multiple bone lesions with cyst formation associated with cutaneous pigmen-

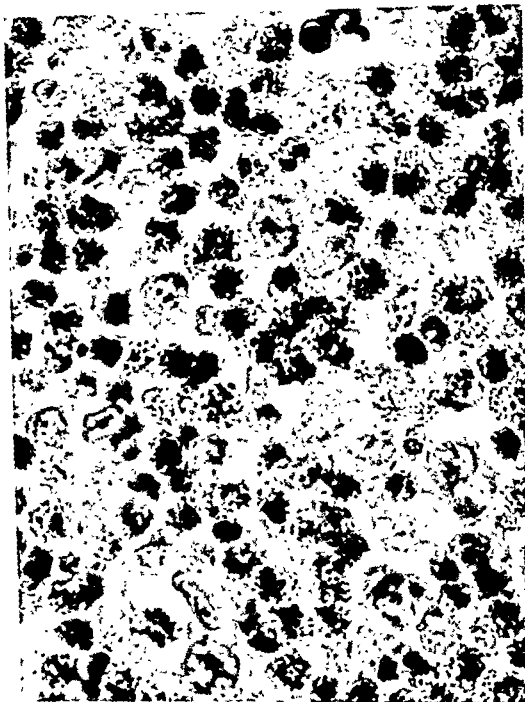


Figure 285. EMB × 400 Photomicrograph of section through soft vascular eosinophilic granuloma of the skull. Note the preponderance of eosinophils in this densely cellular field.

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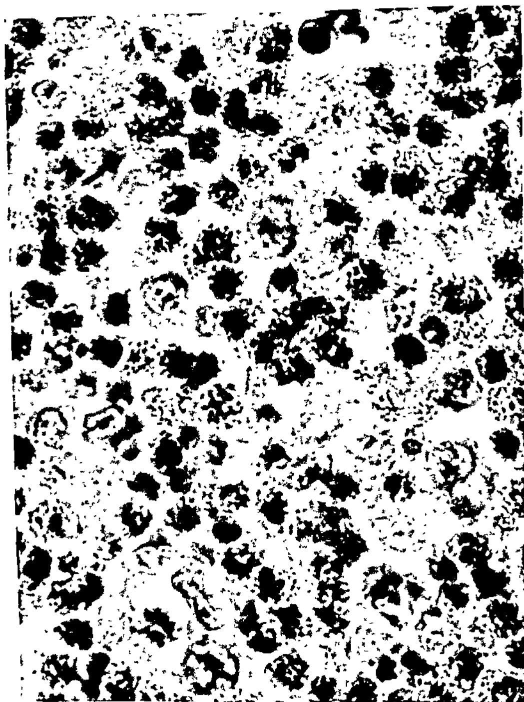


Figure 285. EMB x 400 Photomicrograph of section through soft vascular eosinophilic granuloma of the skull Note the preponderance of eosinophils in this densely cellular field.

tation and sexual precocity in females, the term Albright's syndrome is used.[2]

The skull is frequently involved in fibrous dysplasia.[203] It may be the only part of the skeleton afflicted; under such circumstances there are usually no soft tissue manifestations of the disease. Calcium, phosphorus, acid and alkaline phosphatase and other blood chemical studies are usually entirely normal.[68]



Figure 286. Ten year old boy with right exophthalmos [illegible] temp[illegible] ing th[illegible] [illegible] of the optic foramen

Fibrous dysplasia of the skull alone is commonly first noted during the second half of the first decade of life. The presenting complaint is apt to be cranial deformity or unilateral ptosis or exophthalmos. Deformity is commonly noted as swelling in the supra-orbital or anterior temporal region. It may extend to the facial bones as well as causing prominence of the malar area (Figure 286). This deformity may increase so slowly that its actual presence is long in doubt in the minds of the child's parents. In more severe cases, narrowing of the palpebral fissure, displacement of the eye-ball and impairment of visual acuity become apparent. It is under such circumstances that this disease becomes of neurosurgical significance in childhood.

The x-ray appearance is characteristic in children. The cyst-like, circumscribed areas of diminished density found elsewhere in the skeleton and in the skull of older patients are not ordinarily seen. The commoner picture in childhood is that of extensive new bone formation with marked thickening and increase in density of the involved bones. The base of the skull and adjacent facial bones are most commonly involved. The wings of the sphenoid bone, roof of the orbit, malar bones and lower part of the frontal bone have been the commonest sites in our experience. The process is usually either entirely or predominantly unilateral (Figure 287). Films of the orbital foramina are important in determination of possible compression of the optic nerves. Serial x-ray examination at regular

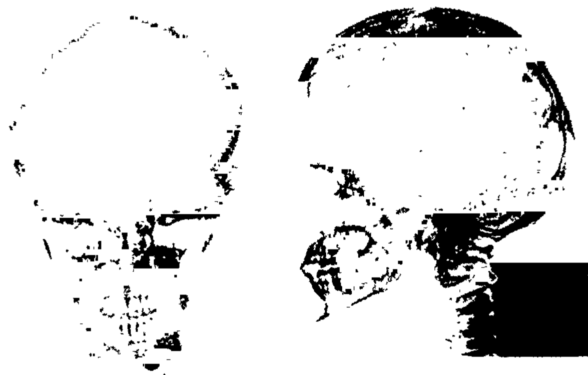


Figure 287. Posterior-anterior and lateral roentgenograms of 10 year old boy with fibrous dysplasia of the skull. Note the characteristic unilateral distribution involving the frontal, orbital and sphenoid bones with encroachment on the optic foramen. In this patient decompression of the optic nerve was carried out because of progressive enlargement of the blind spot and diminution of visual acuity. Same patient shown in Figure 286 and 288.

intervals constitutes the most satisfactory method of following patients during the period of active skeletal growth.

Morbid Anatomy

The fundamental abnormality in this disease is an accumulation of fibrous connective tissue within bone. In the type of fibrous dysplasia affecting the bones at the base of the skull in childhood with which we are concerned here, there is marked thickening due to new bone formation. This bone is very dense yet highly vascular.

Microscopically, the picture is that of cellular, dense fibrous tissue arranged in sheets and whorls. Osteoclastic and osteoblastic activity are seen in the trabeculae of immature bone (Figure 288).

Treatment

The only indication for surgical therapy during childhood is progressive deforming exophthalmos or loss of visual acuity due to compression of the optic nerve. Once the diagnosis has been made, it is important to follow these children by periodic clinical and x-ray examinations. This should

include tests of visual acuity and visual fields. Progressive enlargement of *the blind spot may be the first clue to compression of the optic* nerve within the bony canal.

Surgical treatment for progressive exophthalmos consists in removal of the roof and upper portion of the lateral wall of the orbit exposed by low-placed frontal craniotomy (Figure 305). If there is diminished visual acuity and x-ray evidence of narrowing of the optic canal, the lesser wing of the sphenoid must be removed medially and posteriorly until the optic canal is completely unroofed and the nerve decompressed.

Such surgical treatment, though rarely necessary, is usually quite satisfactory in alleviating exophthalmos and restoring or at least preventing further loss of vision. Once the period of active skeletal growth is completed there is not apt to be further progression of the local lesion.

Surgical treatment for cosmetic reasons alone has not been performed in this clinic and would seem to be contra-indicated except perhaps in extremely severe deformity of the fiotal bone. Large cystic lesions of the cranial vault necessitating excision and cranioplasty have not been seen in young children.



Figure 288 Fibrous dysplasia of the skull. *Note the dense fibrous tissue interlacing between* bone trabecula Specimen from the lesser sphenoid wing of 10 year old boy.



Figure 289A (*Left*). Sixteen months old infant with large osteoma of the frontal and parietal bones.

B (*Right*). Lateral roentgenogram of the skull in this patient. Microscopically, the lesion extends usually well beyond the limits seen by x-ray examination.

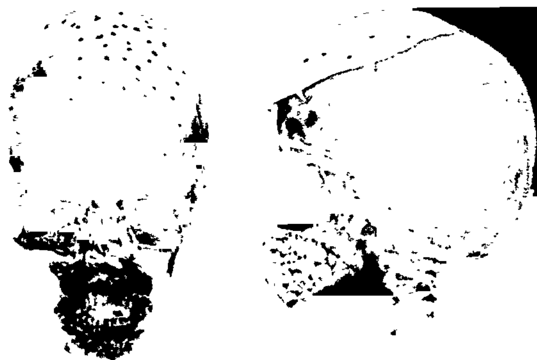


Figure 290. Antero-posterior and lateral roentgenograms of six year old child following tantalum cranioplasty after removal of a large osteoma involving the frontal and parietal bones.

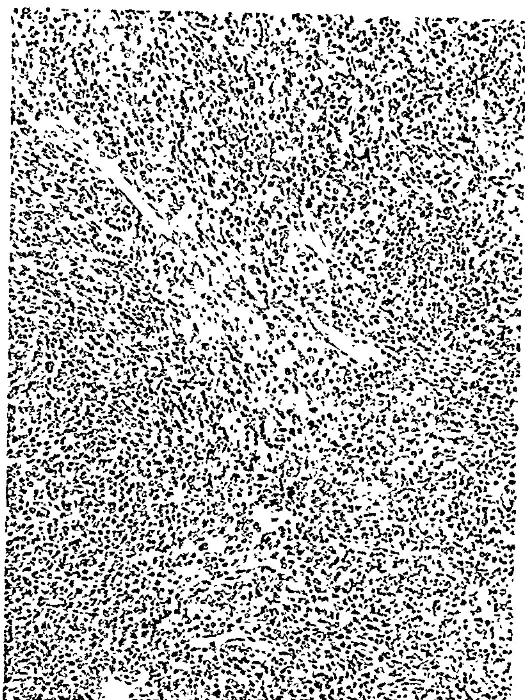


Figure 291. PTAH x 200 Photomicrograph of fibrosarcoma of bone in the occipito-temporal region of a 14 month old boy. Tumor consists of sheets of anaplastic closely packed cells with little stroma. There are many mitoses and atypical cells. The tumor cells are darkly-staining and vary considerably in size and shape. There is a fine collagenous reticulum Numerous thin-walled blood vessels are scattered throughout the tumor

OSTEOMA

Osteoma is a benign, slow growing osseous tumor which rarely involves the skull in young children. This tumor produces overgrowth of bone without destruction and does not invade soft parts. It may however grow to enormous size. The margins of the tumor are difficult to determine both by x-ray and by gross observation. Microscopically, there is predominance of osteoblastic activity with no destruction and no evidence of abnormal collections of cells.



Figure 292. Basal roentgenogram of the skull of a 14 month old boy showing large area of destruction in the region of the petrous portion of the temporal bone on the right due to a primary fibrosarcoma in this area.

The tumor makes its presence known by visible cranial deformity (Figure 289A). The overlying soft tissues are normal but displaced by the bony prominence. Since the tumor apparently arises near the periosteum, where osteoblastic activity is greatest, the major part of the growth is outward. However, there may occasionally be significant displacement of the dura due to growth inward from the inner table of the skull.

By x-ray the bone is thickened and there is increased density. Lesions of the cranial vault are not sharply circumscribed (Figure 289B.) There are no areas of radiotranslucency.

Treatment consists in block resection of this area of bone followed by suitable cranioplasty (p. 173) (Figure 290). The prognosis is excellent.

SARCOMA

Primary malignant sarcomas of bone arising in the cranial vault are rare in childhood. Histologically they may be osteogenic or more commonly simply rapidly growing fibrosarcoma (Figure 291). There is usually an associated soft tissue swelling, apt to be firm and tender and rapidly increasing in size. On x-ray examination, sarcoma of the skull appears as an



Figure 293. Antero-posterior roentgenogram of the chest of the patient shown in Figure 292 six months following radical removal of the skull tumor. At post-mortem there were extensive metastases throughout both lungs and pleura.

irregular area of decreased density with evidence of active bone destruction at the margins (Figure 292).

The course of these lesions is usually extremely rapid. The earliest possible radical surgical excision is the treatment of choice but is usually unsuccessful due to hematogenous spread, principally to the lungs (Figure 293).

TUMORS OF THE MENINGES

The most frequent neoplasm arising from the meninges is the meningioma. As pointed out in the introduction to the section on intracranial tumors (p. 222), however, in infancy and early childhood this tumor, so common in adults, is extremely rare. In our clinic, among 313 intracranial tumors there is only one meningioma in a child under 12 years of age. This was a large parasagittal tumor in the frontal region with invasion of the frontal bone in a child of seven (Figure 294). This tumor had been entirely asymptomatic and was discovered only when a small swelling of the fore-

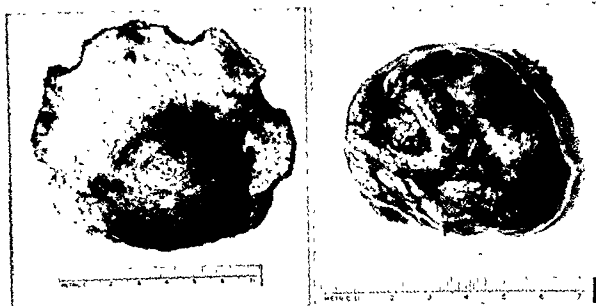


Figure 294. Parasagittal meningioma invading bone in seven year old boy.

A. Inner surface of frontal bone showing invasion of tumor.

B. Tumor mass removed from right frontal fossa together with attached dura and falx.

head was noted by the patient's father. Surgical excision followed by tantalum cranioplasty was satisfactorily carried out. The tumor histologically was a typical benign fibroblastic meningioma (Figure 295).

Two types of more malignant meningeal tumors are occasionally seen. The first of these is leptomeningeal sarcoma. This tumor extends and seeds freely throughout the subarachnoid pathways, primarily about the base of the brain. Symptoms may be convulsive seizures secondary to lesions over the cerebral hemispheres or increased intracranial pressure due to obstruction of the spinal fluid circulation, usually in the cisterna magna or basilar cisternae. Surgical treatment is of no avail. Deep x-ray therapy to the entire subarachnoid pathway is indicated to attempt transient relief of symptoms. Malignant melanoma has been seen involving the meninges diffusely and causing death from obstruction to spinal fluid circulation (Figure 296).

The second type of malignant meningeal tumor is fibrosarcoma of the dura (Figure 297). These tumors are primary in the dura and do not invade the underlying leptomeninges or brain. They are similar to tumors of collagenous connective tissue seen elsewhere in the body (Figure 298). The tumor may be diffuse, bilateral and sacular with spaces containing semi-gelatinous material and lined by solid neoplasm. It may occur as a single, solid fibrous lesion producing symptoms and signs consistent with any other type of local mass in this region. In the latter type total surgical removal may be feasible. In inoperable lesions, x-ray therapy has been

reported to show an immediate strikingly favorable response followed by recurrence and death.[8]

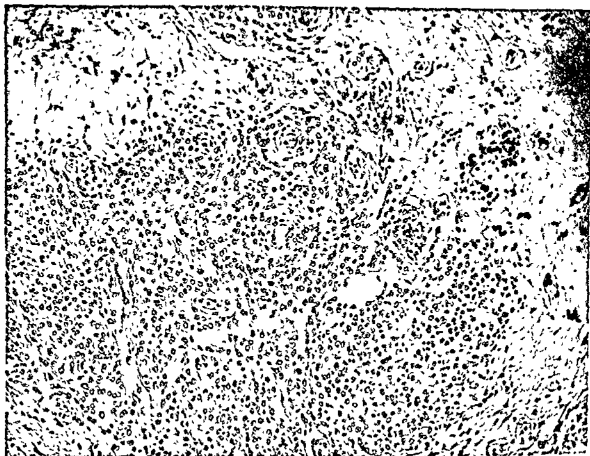


Figure 295. PTAH x 200 Photomicrograph of meningioma removed from right fronto-parasagittal region. The tumor cells are arranged in sheets and characteristic whorls The cells have round to oval nuclei and a moderate amount of cytoplasm The wrapping tendency of the tumor cells characteristic of fibroblastic meningiomas is well shown.

Figure 296. Two months old infant with extreme hydrocephalus due to extensive involvement of the meninges as well as the skin by malignant melanoma.

Figure 297. Five years and four months old child with extensive fibrosarcoma of the dura showing protrusion through the skull.

extra-ocular muscles, so that subjective diplopia and objective strabismus occur.

Unilateral loss of visual acuity is seldom an early complaint among infants and children. Frequently it is never suspected by the parents and is discovered only during testing as a result of some other primary complaint. Failure of a pupil to react to light does not occur until the loss of vision is profound. Unilateral fixed dilatation of the pupil is therefore more apt to be associated with impairment of third nerve function than it is with diminished light perception. The two can be differentiated, of course, by testing the consensual reaction to light.



Figure 299. Four months old infant with left retro-orbital pseudotumor or chronic myositis of the extra-ocular musculature of unknown etiology. Note slight exophthalmos and narrowed palpebral fissure on the left.

Occasionally a mass may be palpable adjacent to the eye-ball causing its displacement. Dilatation of scleral and conjunctival vessels may occur as the result of impaired venous blood flow from the globe.

Funduscopic examination may give the first suggestion of an orbital lesion by demonstrating (1) unilateral papilledema, as in the presence of a mass producing obstruction to venous return from the retina; or (2) by demonstrating unilateral primary optic atrophy due to either an infiltrating or extrinsic lesion of the optic nerve which impairs its function without causing venous obstruction.

Clinical examination should include:

1) Measurement of the diameter of both pupils and estimation of their comparative reactivity to light and accommodation.

2) Determination of visual acuity in each eye. This is possible to a gross degree in any infant old enough to follow light and in more detail in children old enough to recognize letters, numbers, or small pictures of familiar objects.

3) Observation of extra-ocular movements.

4) Funduscopic examination.



Figure 300 (*Left*). Six year old boy with ptosis of the left upper eye-lid. Left pupil is slightly dilated but vision normal. Skull x-rays and arteriogram negative. Patient subsequently developed slight exophthalmos. Exploration of left orbit through left frontal craniotomy revealed rhabdomyosarcoma, primary in the orbit.

Figure 301 (*Right*). Ten year old girl with right exophthalmos and displacement of the globe downward and outward due to neurofibroma of the optic chiasm extending into the orbit. Vision markedly impaired. Note the small cafe-au-lait spot in the right supraorbital region.

5) Exophthalmometry.

6) Determination of visual fields by perimetry or tangent screen test in children old enough to cooperate and by repeated confrontation with desirable objects in young children.

7) Auscultation for bruit over the eye-ball.

Because of frequent metastases to the orbit from neuroblastoma, careful palpation of the abdomen should be performed in every child with unilateral exophthalmos or other evidence of an orbital mass. This should be supplemented by intravenous urography due to the frequent suprarenal location of the primary tumor (Figure 302).

The entire skin area should be examined carefully for cafe-au-lait spots since optic nerve lesions are occasionally associated with generalized neurofibromatosis (Figure 303).



Figure 302. Intravenous urogram of 14 month old male infant with abdominal neuroblastoma displacing left kidney downward and right kidney laterally. Patient had metastasis to the right orbit causing exophthalmos.

X-ray Examination

Roentgen study of these children should include films of the skull, orbits, chest and abdomen. Occasionally, survey films of all the long bones and intravenous urograms are also indicated. Routine skull films may show thinning or destruction of the roof or lateral wall of the orbit or of the lesser wing of the sphenoid bone; they may show deformity of the sphenoid fissure. Oblique films of the orbits are examined for comparison of the contour and size of the optic foramina. The latter are enlarged with infiltrative tumors of the optic nerve (Figure 304) and may be smaller than normal in polyostotic fibrous dysplasia involving the orbital bones.

Completely normal x-ray films of the skull and orbits do not rule out orbital tumors since these lesions may grow to considerable size without encroaching upon the optic canal or orbital walls.

Treatment

Occasionally, anterior exploration of the orbit may be satisfactory for exposure and biopsy of an anteriorly placed neoplasm, particularly when this is visible or palpable pre-operatively. However, in most instances the safer and more satisfactory method of exposure of orbital lesions and certainly the preferred exposure where radical excision is planned is by means of transfrontal craniotomy.

In infants and children a small frontal bone flap placed very low and raised laterally on a hinge of temporal muscle is used (Figure 305). Since the frontal sinus is usually undeveloped in this age group it is easy to place the bone flap just above the orbital ridge. A coronal scalp incision just behind the hair-line is recommended in children since the forehead is easily reflected, permitting a low bone flap and since it heals rapidly and satisfactorily without visible scar formation.

The dura is separated from the roof of the orbit, which forms most of the floor of the frontal fossa. Withdrawal of spinal fluid through a lumbar puncture needle facilitates retraction of the brain for this exposure. The roof of the orbit is penetrated and sufficient bone removed with rongeurs from the roof, lateral wall and wing of the sphenoid bone to expose the orbital contents. The bony removal may be

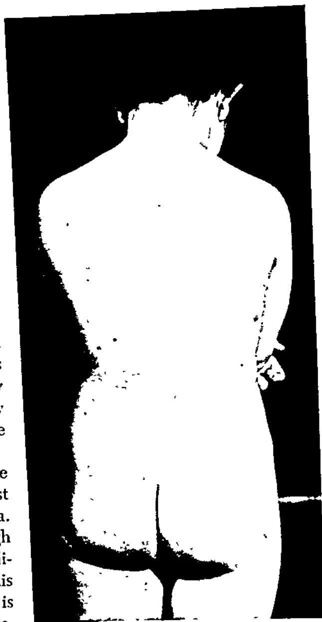


Figure 303 Posterior view of the trunk of a 10 year old girl showing numerous cafe-au-lait spots characteristic of Von Recklinghausen's neurofibromatosis. This patient had extensive neurofibromas about the optic chiasm. Same patient shown in Figure 301.



Figure 304. Film of the right optic foramen showing extreme enlargement in a 10 year old girl with Von Recklinghausen's disease and a neurofibroma involving the optic chiasm. Patient had right exophthalmos and was blind in this eye.

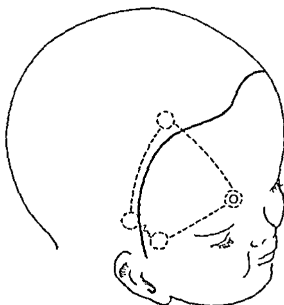


Figure 305. Plan for exposure of parapituitary area or orbital exploration. Heavy line indicates position of the skin incision just behind the hair-line. Dotted lines indicate position of the bone flap which is broken laterally on a temporal hinge. A trephine button is removed medially and inferiorly so that it can be replaced at the end of operation.

safely extended through this approach to explore the superior orbital fissure or to unroof the entire optic canal. If necessary, the dura is opened to expose the proximal portion of the optic nerve and the chiasm and to perform combined intra- and extradural resection of optic nerve tumors.

After removing the roof of the orbit, the capsule should be incised parallel to the long axis of the orbit taking care not to injure the ophthalmic branch of the trigeminal nerve or the nerve to the levator palpebrae muscle. The capsule should be opened widely before exploration of the soft tissues is begun.

If there has been considerable handling of orbital tissues at the time of operation in a child who already exhibited a moderate degree of exophthalmos, it is wise to perform temporary canthorrhaphy at the time of craniotomy. This is accomplished by placement of two or three mattress sutures of silk through the lid margins approximating them gently but not tightly so as to cover the globe completely. These sutures can be released usually in five to eight days with safety.



Figure 306 Firm rubbery swelling in the outer supraorbital area of 10 months old infant with unilateral ptosis and exophthalmos due to metastatic neuroblastoma.



Figure 307. Low-power photomicrograph of metastatic neuroblastoma to the skull and orbit. Primary tumor in the region of the left adrenal gland. Note the masses of densely staining small cells with tendency toward rosette formation.

Morbid Anatomy

The gross appearance of orbital tumors varies widely at the time of operation. Metastatic tumors, such as neuroblastoma (Figure 306) and lymphoblastoma, are apt to be very cellular, vascular, purplish-pink, friable masses of tissue easily distinguishable but not discretely separable from the orbital soft tissues. Their histological picture is comparable to these tumors elsewhere in the body (Figure 307). Fibrosarcomas or rhabdomyosarcomas are tougher, less vascular, adherent to orbital tissues and often giving a false appearance of complete encapsulation. Gliomas of the optic nerve may appear as nodular, rubbery, pinkish-gray tumor masses deep in the orbital soft tissues, or simply as pinkish, edematous diffuse enlargement of the optic nerve itself. For the most part these are mixed tumors histologically, the commonest of all patterns being those of astrocytoma or spongioblastoma polare (Figure 308). Neurofibromas are well encapsulated, firm and adherent to the nerve sheath (Figure 309). Hemangiomas are usually



Figure 308. PTAH x 200 Photomicrograph of section through glioma of the optic nerve in a five year old boy. Note interlacing neurofibrils. Predominant cell is an astrocyte. Tumor is relatively avascular.

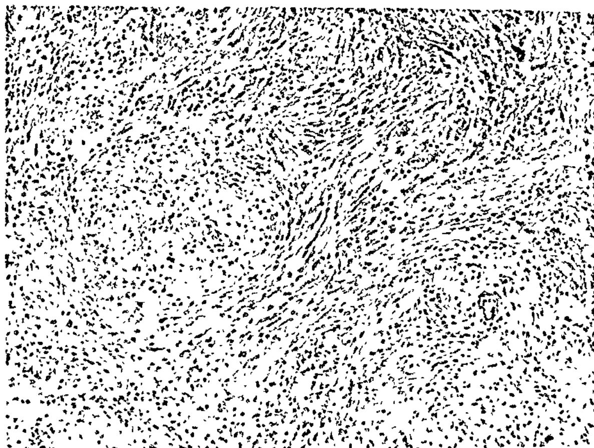


Figure 309. TBE x 200 Photomicrograph of neurofibroma removed from the region of the optic chiasm bilaterally in a 10 year old girl with Von Recklinghausen's neurofibromatosis. Note whorls and palisades of spindle shaped cells coursing in the plane of the section and perpendicular to it. The cells have ovoid nuclei and scanty cytoplasm tapering at each end. The tumor is relatively avascular.

readily recognizable as tangled masses of vessels of varying size. Retrobulbar pseudotumor is distinguished by diffuse infiltration, edema and pallor of the orbital fat and muscles giving the impression of marked increase in pressure of the entire orbital contents (Figure 310).

Results

The results of treatment of optic nerve gliomas vary naturally with the extent of the lesion at the time of surgery. If there is diffuse extension from one optic nerve into the chiasm or into the other optic nerve, radical excision is obviously undesirable (Figure 311). Several long-term results have been reported following biopsy and deep roentgen therapy.[162] Rarely, a glioma may be limited to one optic nerve so that resection proximal to the tumor but distal to the optic chiasm with enucleation of the eye may be expected to result in permanent cure. When biopsy indicates neuroblastoma or lymphoblastoma, there is little virtue in attempting radical local removal. Deep roentgen therapy is indicated to the orbit in such a fashion as to try

and protect the globe itself. Although temporary remission has occurred with this treatment, eventual fatality is the rule.

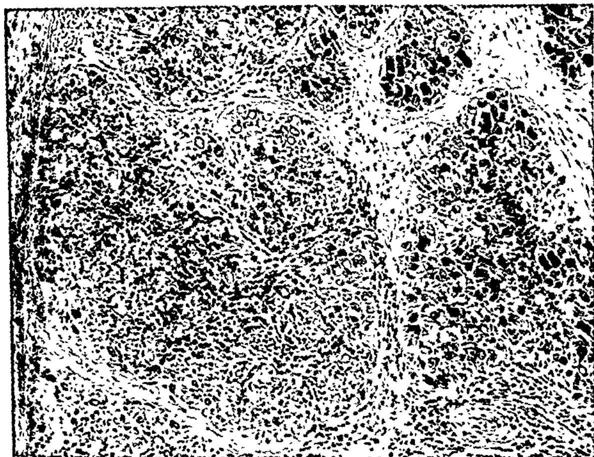


Figure 310. Low-power photomicrograph of chronic myositis of the extra-ocular muscles producing unilateral exophthalamos in a 6 9/12 year old girl. Note diffuse infiltration of muscle bundles by numerous chronic inflammatory cells. There is also separation of the muscle bundles by edema.

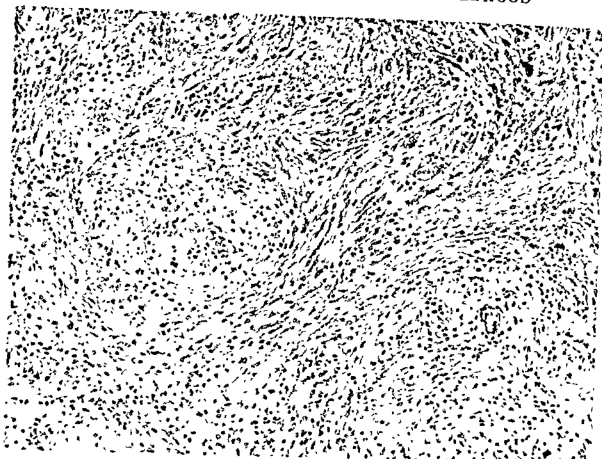


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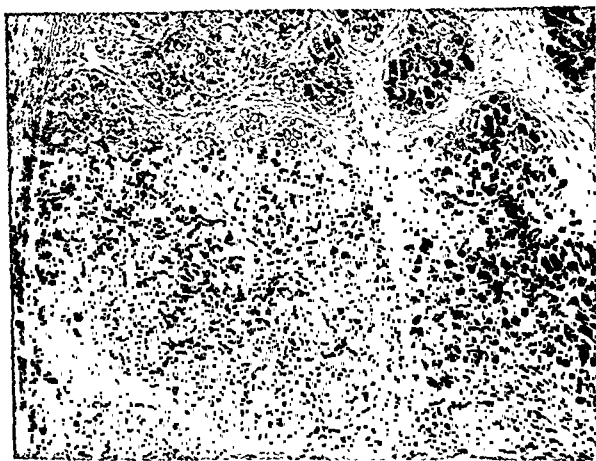


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Figure 311. Two years and four months old boy showing marked left exophthalmos and partial ophthalmoplegia due to glioma of the optic chiasm. Tumor involved both optic nerves and eventually proved fatal.

PART V

INTRASPINAL TUMORS

extensions of intracranial neoplasms. In 1944[82] he reported an additional 114 cases collected from 1933 to 1942. From our clinic, 16 children with intraspinal tumors were reported in 1938.[89] Between 1938 and 1952, 47 additional patients with spinal tumors have been treated, or a total of 63 patients under 12 years of age. During the same interval, 313 infants and children with intracranial tumors were treated. It is apparent, therefore, that neoplasms are only about one-fifth as common within the spinal canal as within the intracranial chamber during childhood.

The incidence of various tumor types in childhood varies considerably from that of adult life. In any group of adult cases of intraspinal tumor, a large percentage are derived from the meninges or nerve roots. These are almost completely absent in childhood. Instead, in addition to intrinsic gliomas of the spinal cord, many so-called "congenital" tumors are discovered in early life. These include teratomas, teratoid tumors and dermoid cysts. In this discussion are included only patients with a discrete tumor

INTRASPINAL TUMORS

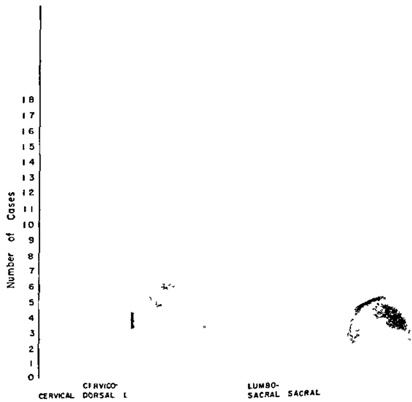


Figure 313. Distribı ... 'nal tumors ' 'dhood. The high ... cervical 'ar regions is di ... ..ce of genital

located within the spinal canal in whom symptoms and signs were produced by this tumor mass and not by an associated developmental defect that might have been present. Lipomatous malformations occurring in conjunction with occult spinal defects are not considered here but rather in the section on spina bifida (p. 17). Likewise, spinal epidural cysts, intraspinal meningoceles, inflammatory granulomas and spinal extensions of intracranial tumors (medulloblastoma, ependymoma, astrocytoma) are excluded.

Intraspinal tumors occur throughout the pediatric age period. Our cases varied in age at the time of hospital admission from 10 weeks to 11 10/12 years. The distribution is indicated in Figure 312. According to Hamby's collected figures, the incidence of spinal tumors goes up rather sharply between 13 and 15 years of age.[82] There is no sexual preponderance.

The anatomical distribution of these lesions is of some interest since they appear to be concentrated principally in the cervical and lumbar regions of the spine (Figure 313). This is especially true of the "congenital" tumors, and is consistent with the prevailing anatomical distribution of other types of embryological disorders of the spinal axis (p. 22).



Figure 314. Six year old girl admitted to the hospital with question of poliomyelitis because of weakness of the left leg There was, however, marked pain in the low back and leg and a vague sensory level in the mid-thoracic region. The patient proved to have an astrocytoma of the thoracic cord, partially cystic. Excellent recovery following two-stage operative removal.

Symptoms and Signs

There is perhaps no neurosurgical condition in which the history is so apt to be inadequate, and in which there is such a paucity of reliable physical signs early in the course of the disease. These factors, together with the rarity of intraspinal tumors in childhood, contribute to the frequency with which the diagnosis is long delayed or completely overlooked.

Children with minimal symptoms and signs suggestive of spinal cord compression present extremely difficult diagnostic problems. Pain is not a common symptom since tumors involving primarily the nerve roots and meninges are almost unknown in childhood. When pain is present, the radiating pattern so characteristic

of root compression in an adult is difficult or impossible to determine in a child. Minor degrees of numbness, paresthesia, loss of tactile and thermal acuity, loss of position sense in the lower extremities — all sensory phenomena of which an intelligent adult may be distressingly aware before the examiner can elicit any abnormality objectively — are usually of no avail in analysis of the infant and young child. Similarly, early disturbance of rectal and vesical sphincter control may be impossible to evaluate in the child who is untrained or in the training period.



Figure 315. Three and ten-twelfths years old boy with a neurofibrosarcoma invading the cervical spinal canal and involving the brachial plexus on the left. Note tilting of the head to the left side and limp position of the left upper extremity.

Only too often the first suggestion of an intraspinal disorder comes with parental detection of motor weakness, usually in the lower extremities. This may be noted as a gait disorder or as an inability or unwillingness to bear weight normally. Unfortunately, progression from minor weakness to extensive paraplegia sometimes may be very rapid. Among our patients, by far the commonest initial complaint was motor impairment, recognized as difficulty in walking or bearing weight (Figure 314), as a localized weakness of one upper extremity (Figure 315) or as muscular spasm. Pain in the back, abdomen or thighs was the next commonest complaint. Urinary incontinence was the primary disorder in a few and several were first seen because of some localized abnormality visible in the skin over the spine.



Figure 316. One year and nine months old boy with partial quadriplegia of three weeks' duration due to cystic teratoma of cervical cord. Note accompanying spina bifida occulta and vertebral body defects in this area.

The frequency with which skin malformations occur in association with intraspinal teratomas and dermoid cysts has been stressed elsewhere (p. 69).

It is evident from these observations that accurate early diagnosis of intraspinal tumors in children implies first, realization that such a lesion is a possibility and second, investigation of minimal symptoms and signs by objective diagnostic aids, including spine roentgenograms, spinal fluid studies and contrast myelography.



Figure 317 Extreme antero-posterior widening of lumbo-sacral canal with concavity of posterior surface of vertebral bodies and destruction of laminae and pedicles due to huge intraspinal teratoma in a 1 10/12 year old infant.

There are many pitfalls in the differential diagnosis of spinal disorders in infancy. Weakness of all four extremities as well as trunk musculature incident to a cervical cord tumor may be misinterpreted as due to general prostration and malnutrition. Elevation of temperature and paralysis of intercostal muscles may lead to a diagnosis of pneumonia rather than cervical cord tumor. Regression in bladder and bowel training may be misinterpreted as a behavior problem rather than the earliest evidence of cord compression. The onset of motor weakness in a child living in a community where anterior poliomyelitis is epidemic frequently leads to unwarranted diagnosis of the latter condition. This is enhanced by the difficulty in eliciting objective sensory changes in young children. However, with time, patience, a little experience and repeated observations, a careful sensory examination can usually be performed on even the youngest and most irritable infant.

The possibility of intraspinal tumor should be considered whenever tentative diagnoses are entertained of cerebral palsy, anterior poliomyelitis, spina bifida occulta with neurological involvement, hereditary muscular dystrophy, or birth injury to the spinal cord. Under such circumstances, if compression of the spinal cord is a possibility, complete sensory, motor

and reflex examination must always be supplemented by spinal fluid and x-ray studies.

Spinal Fluid Findings

Probably the most important diagnostic procedure in determining the presence of intraspinal tumors is performance of a careful lumbar puncture with examination of the spinal fluid. If necessary, this should be done under sedation or even full anesthesia. In our experience with children this has invariably revealed abnormal findings in the presence of an intraspinal mass lesion. Occasionally the tumor type as well as its presence may be diagnosed at this time. Gelatinous material may be aspirated directly from a teratomatous cyst. Identification of squamous epithelial cells in the spinal fluid may indicate the presence of a dermoid cyst. Inability to obtain any spinal fluid at all at several lumbar interspaces may indicate an extensive cauda equina tumor such as an ependymoma.

The spinal fluid pressure itself is relatively unimportant. It is usually normal or low. Little or no fluid may be obtained. Spinal fluid dynamics may reveal evidence of a partial or complete block of the subarachnoid space. However, normal dynamics do not, of course, rule out tumor. Existence of a block always indicates surgical exploration.



Figure 318. Antero-posterior and lateral roentgenograms of the spine of seven and a half years old boy with dermoid cyst of the conus medullaris. Note widening of spinal canal at T_{11} and L_{1} with thinning of pedicles and scalloping of vertebral bodies.

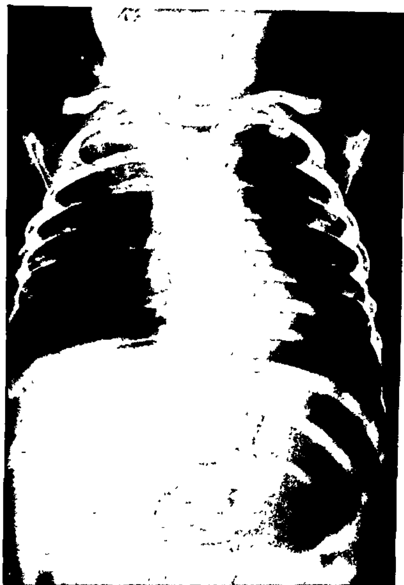


Figure 319. Antero-posterior roentgenogram of the spine of an 18 month old girl with neuroblastoma causing paraplegia. Note widening of the spinal canal at T_1 to T_3 with extraspinal soft tissue mass at the right apex.

Because of the *high frequency of the intradural location of spinal* tumors in childhood, some degree of xanthochromia of the fluid is common and an elevation of the total protein content is the rule. In our patients the latter has varied from 50 mgms. per cent up to as high as 3.95 gms. per cent. With marked xanthochromia and elevation of protein content in the presence of a complete block, the fluid frequently clots on standing.



Figure 320. Collapse of body of L_1 associated with sudden onset of paraplegia at 11 months of age due to metastatic papillary adenocarcinoma. Sacro-coccygeal teratoma had been removed shortly after birth.

X-ray Findings

In every child with a suspected lesion of the spinal cord or cauda equina, plain antero-posterior and lateral roentgenograms of the entire spine should be made. Additional oblique, spot and other special films are then made as indicated. Abnormalities of the vertebral column adjacent to "congenital" intraspinal tumors are extremely common. These include spina bifida, hemivertebrae, abnormally fused vertebrae, and abnormal pedicles or facets (Figure 316). A slowly expanding intraspinal mass may result in widening of the canal, thinning of the facets, or increased concavity of the posterior surface of the vertebral bodies (Figures 317 and 318). More rapidly growing extradural lesions may show destruction of the pedicles and laminae without any evident widening of the spinal canal. Intraspinal tumors with extraspinal soft tissue extensions may sometimes be detected by x-ray (Figure 319). Metastatic tumor to the spine may result in paraplegia secondary to sudden collapse of a vertebral body with acute angulation of the cord; this is readily visualized in a lateral x-ray film (Figure 320).

Contrast myelography with pantopaque is a helpful adjunct to tumor localization. This procedure can be carried out readily and safely in infants or children of any age. It should always be performed under fluoroscopy so that the flow of opaque medium is visualized directly. In the absence of changes in the plain roentgenograms, and with poor localization clinically

in an uncooperative child or small infant, determination of the correct level of the lesion by myelography is invaluable in planning surgical treatment (Figure 321). In the presence of a block, pantopaque may be introduced in the lumbar region to demonstrate the lower extent of the lesion (Figure 322) or in the cisterna magna to demonstrate the upper extent.

Morbid Anatomy

The types of spinal tumors seen among children in our clinic are indicated in Figure 323. In general, they fall into three groups:

(1) "Congenital" tumors, ranging from simple dermoid cysts at one end of the scale of increasing complexity to the highly varied tridermal teratomas at the other.

(2) Intramedullary gliomas, including astrocytoma, ependymoma, medulloblastoma and glioblastoma multiforme; and

(3) Extradural extensions of paraspinal lesions, including neuroblastoma and reticulum cell sarcoma and lymphosarcoma. A variety of rarer lesions may also be encountered.

In all instances, the "congenital" tumors have been in part cystic.[20, 24,



Figure 321. Pantopaque myelogram in 1 10/12 year old infant with intramedullary astrocytoma extending from C_4 to T_3 The opaque medium outlines the upper and lower borders of the lesion accurately

79, 92, 159 The cysts may be single or multiple and may vary in size, from almost microscopic structures to large lesions lying in the subdural space or extending throughout several segments of the spinal cord. The cyst walls may be thin and parchment-like or thick and resistant. In the latter instance the cyst wall is usually a mixture of tumor and reactive gliosis of the adjacent tissue.

The fluid in these cysts varies greatly. It may be a pale, canary yellow clear fluid which clots on standing (Figure 324); it may be cloudy because of contained cells, or deeply amber or chocolate colored because of recent hemorrhage; it may be opaque gelatinous fluid from a cyst lined by mucin secreting cells; it may contain keratinized cells, sebaceous debris and hair when the lining is stratified squamous epithelium. Among the teratomas, of course, the histological possibilities of the solid portions of the tumor



Figure 322. Spot film during pantopaque myelography in seven year old girl showing opaque medium completely blocked in upward ascent at T_6 by intraspinal mass which proved to be an extradural reticulum cell sarcoma. Soft tissue extraspinal mass is also well shown.

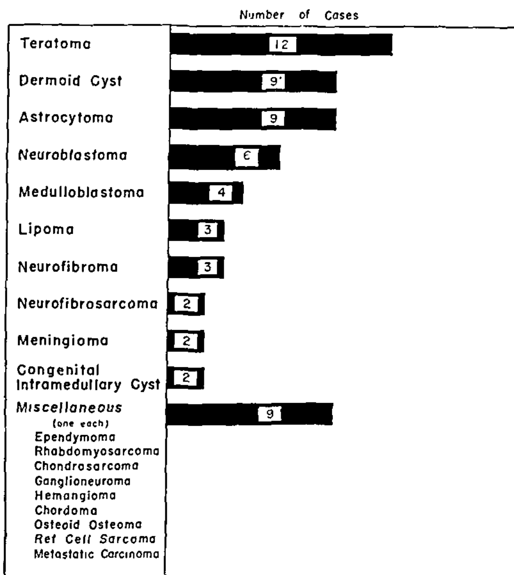


Figure 323. Distribution of tumor types in intraspinal tumors in children. The striking difference from adult patients is the preponderance of congenital tumors and the rarity of meningiomas and neurofibromas.

are many (Figure 325). There is always the chance of malignant change occurring in one of the tissue types at any time.

There is need for little comment regarding the intramedullary gliomas. The astrocytomas have frequently been partially cystic; the ependymomas usually solid. All of these tumors are poorly demarcated from normal neural structures and difficult to differentiate from reactive gliosis (Figure 326). Their histological features have been similar to those of the intracranial gliomas (Figure 327).

Neuroblastoma, one of the common malignant tumors of childhood, is primary in the paraganglionic areas but frequently invades the spinal canal through one or more intervertebral foramina, extending as a cuff around the nerve root. Intraspinally, it grows up and down in the extradural space and by simple displacement may compress the contents of the dura rapidly and severely. It is an extremely vascular, usually quite soft, cellular tumor crossed with occasional fibrous tissue septa (Figure 328). It never penetrates the dura or nerve sheaths. Sarcomas, primary in extraspinal lymphoid tissue, behave in essentially the same manner as neuroblastoma when they invade the spinal canal.

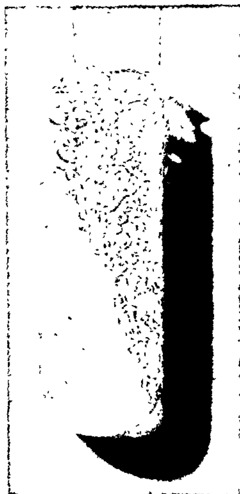


Figure 324. Clear, xanthochromic high protein fluid removed from cystic teratoma of the spinal cord. Note the large bubbles which formed on the fluid when it is shaken in a test tube.

Treatment

The optimum treatment of tumors within the spinal canal is surgical excision after accurate localization. There is no need here to elaborate on the technique of laminectomy. In infants and children a comparatively greater exposure is usually made than in adults. Operations are ordinarily performed under general endotracheal anesthesia with the patient in the prone position. If the cervical region is to be approached, a horseshoe shaped cerebellar type of head rest is used. The operative area is generously prepared and draped no matter what the limits of the original incision, as often these tumors extend for a considerable distance within the spinal canal. Usually, the caudal margin of the tumor is first exposed and laminectomy then carried cephalad as far as necessary for satisfactory exposure.

In infants and young children laminectomy of several segments is apt to be a more shocking procedure than seems readily apparent at the time because of the comparatively small size of the incision. It is therefore wise to anticipate difficulty by having a constant intravenous catheter or cannula in place for infusion of fluids and blood throughout operation. Adequate elevation of the shoulders and hips from the operating table to permit good respiratory exchange is vital. In cervical spine surgery, facilities for maintenance of positive pressure respiration should always be available.

In general, standard neurosurgical methods are appropriate to the removal of spinal cord tumors in childhood. Two considerations, however, which warrant special discussion are first, the conservative subtotal excision of dermoid and teratomatous lesions attached to the cord, and secondly, the advantage of two-stage excision for many intramedullary gliomas.

The so-called "congenital" tumors may be extradural, subdural or within or closely attached to the spinal cord or roots of the *cauda equina* (Figure 329). Complete excision of lesions in the extradural and subdural



Figure 325. Photomicrograph through solid portion of cystic teratoma removed from lower thoracic spinal canal of 5 2/12 year old boy. Note epithelial, fibrous and glandular tissue elements.

space is usually possible with no particular difficulty or untoward sequellae. When these benign lesions are attached closely to the cord, however, radical excision is often impossible and usually the hazards of attempting complete removal are not warranted.[92] Simple evacuation of accessible cystic structures and removal of whatever cyst wall or solid tumor can be freely mobilized is the method of choice. This conservative type of excision has been followed repeatedly by marked and prolonged improvement (Figure 330). In spite of the discouraging degree of compression of the spinal cord apparent at the end of operation in many instances, virtually complete recovery of function has occurred. On the other hand, attempts to remove cyst wall or solid tumor which is closely adherent to nervous tissue and separable only by sharp dissection will almost invariably result in an increased neurological deficit which may be permanent.



Figure 326. Intramedullary cystic astrocytoma of the lower cervical and upper thoracic spinal cord in a 6 9/12 year old child exposed at operation. Note the pale color and enlargement of the spinal cord throughout the upper two-thirds of the exposure.

Gliomas within the spinal cord may well be approached by a planned two-stage procedure. At the first operation the involved cord is exposed and a mid-line dorsal incision made in the cord over the lesion. A small biopsy can be taken at this time for histological identificaton. The dura is then left open and a thin layer of gel-foam placed over the tumor. The rest of the wound is closed tightly. When the wound is reopened in five to eight days the tumor usually will have demarcated itself from the surrounding cord and may sometimes be almost entirely extruded from the cord, thus making radical removal possible with minimal damage to normal structures (Figure 331). The original dorsal incision in the cord should extend the full length of the tumor.

Another problem in surgical treatment which deserves comment is the management of intraspinal neuroblastoma. When signs of cord compression exist, particularly if they are progressing rapidly in severity, it is never justifiable to await the effects of x-ray or chemotherapy. Laminec-



Figure 327 TBE x 200 Photomicrograph of section from intramedullary astrocytoma of the cervical cord in a 22 months old female infant. The tumor is cellular with a dense reticular stroma. Tumor cells have a large pale oval nuclei. Mitotic figures are very rare. Intertwining fibrillary processes are prominent. Cells are evenly distributed throughout the tumor with no tendency towards perivascular or other specific arrangement.

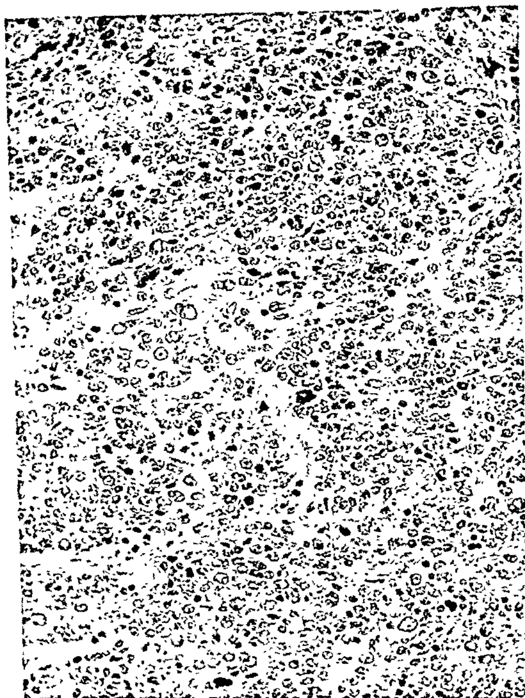


Figure 328 TBE x 400 Photomicrograph through section of extradural neuroblastoma removed from the upper thoracic spine of a 17 months old girl. The tumor consists of irregular masses of cells arranged in groups with a tendency toward rosette formation. There are many mitoses. The neuroblasts have large rounded vesicular nuclei with scant cytoplasm.



Figure 329. Dermoid cyst of the conus medullaris in a seven and a half years old boy exposed at operation. Child's principal complaint was of pain in the lower abdomen for which he had had an interval appendectomy. Note intimate adherence of the tumor to the conus medullaris.

tomy with complete excision of the intraspinal tumor mass should be performed as early as possible. This is followed then with surgical or roentgen therapy of the extraspinal neuroblastoma (Figure 332). This type of treatment with subsequent roentgen therapy has been followed by marked recovery of function, which has persisted in one case as long as 14 years.

Extensive laminectomies in growing children are apt to be followed by skeletal deformities not noted in older patients. In the lumbar region an increased lordosis may ensue, in the dorsal region a kyphoscoliosis (Figure 333), and in the cervical region again an exaggerated lordosis. Ambulatory plaster jackets or braces should therefore be employed usually for at least six to 12 months when several segments have been laminectomized in a young child to minimize this deformity.

Radiation therapy has little application in childhood spinal lesions other than in extradural neuroblastoma and lymphosarcoma. It is also recommended in cases of malignant gliomas incompletely excised.



Figure 330A (*Left*). Two years and four months old infant showing quadriplegia due to cystic teratoma of the cervical spinal cord.

B (*Right*). Same patient at 18 years of age. No evidence of recurrence after simple aspiration of cyst.

(A. reprinted through the courtesy of The Yorke Publishing Company, Inc. from *Am J. Surg*, 39.342-376, 1938.)



Figure 331. Intramedullary astrocytoma of thoracic cord with recent hemorrhage in a 5 11/12 year old girl. Dorsal columns split and biopsy obtained at first stage At second stage six days later, tumor was fairly well demarcated and extruded from cord. Four years after excision patient has only hyperactive reflexes and unsustained ankle clonus, carries on normal activity.

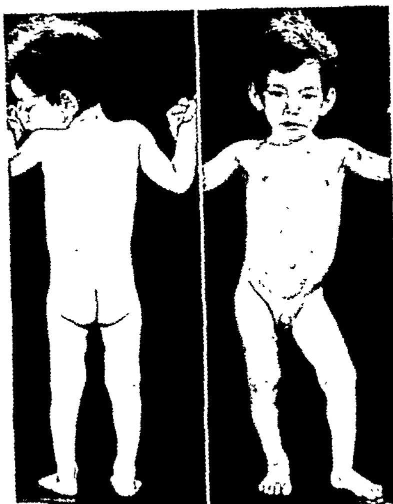


Figure 332. Two years and three months old boy seen because of weakness of legs. Examination also revealed abdominal mass. Photographs show patient after removal of extradural tumor at laminectomy and retroperitoneal abdominal tumor locally and at laparotomy. Patient alive two years after this surgery and x-ray therapy.

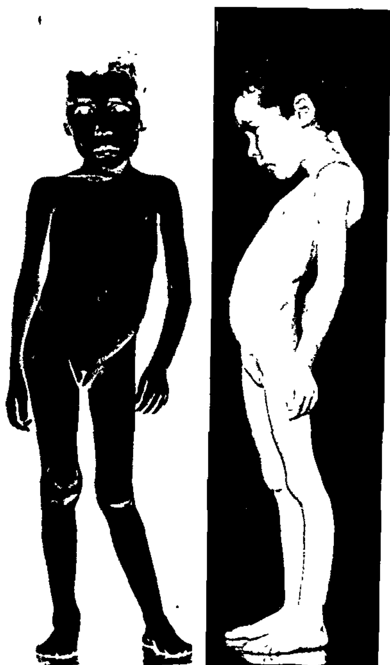


Figure 333. Five and one-twelfth year old girl one year after extensive cervical-thoracic laminectomy for removal of an astrocytoma of the cord, partially cystic. Note the marked degree of kyphoscoliosis which has developed in spite of post-operative splinting.

PART VI

INFECTION

Chapter I

Osteomyelitis

OSTEOMYELITIS of the skull in childhood has almost disappeared within the past several years, principally due to the improved treatment of sinusitis, mastoiditis, and pulmonary suppuration. The experience of The Children's Medical Center in the last five years is limited to rare occurrence of post-operative infection and a single instance of localized osteomyelitis associated with an infected dermoid cyst of the skull (Figure 334). Metastatic osteomyelitis from distant foci of infection and osteomyelitis as a result of direct or venous extension from acute sinusitis or mastoiditis have become almost unknown since the general use of antibiotics in the treatment of primary infections.

In childhood the organism causing bone infection has usually been *staph. aureus* in metastatic lesions and those secondary to infection in the adjacent soft tissues. In lesions due to extension from the air sinuses or mastoid, the *pneumococcus* and *streptococcus* have been more frequently the causative agent, particularly in infants. Osteomyelitis should be suspected in post-traumatic and post-operative infections whenever a draining wound does not heal promptly with proper surgical and antibiotic treatment. The persistence of a chronically draining sinus even though cultures are negative is always indication for careful x-ray examination of the underlying bone. In such patients, particularly when they



Figure 334. Local osteomyelitis of frontal bone associated with infected dumbell-shaped intra- and extracranial dermoid cyst.

have been on antibiotic treatment, there may be no fever, leucocytosis or other systemic evidence of infection.

The cardinal local signs of acute osteomyelitis of the skull are edema, tenderness, redness, and perhaps increased warmth of the overlying scalp. Signs of associated meningeal infection are rare since the dura is an excellent barrier to the spread of sepsis from without. In severe, fulminating osteomyelitis there may be high, spiking fever, shaking chills, leucocytosis up to 20,000 to 30,000 w.b.c./cu. mm. and extreme prostration.

The diagnosis of osteomyelitis is usually confirmed by x-ray examination. In young children this may be difficult because minor degrees of bone destruction are difficult to determine with certainty, particularly in the presence of much edema in the soft tissues. Characteristically, irregular, patchy areas of decreased density, with indistinct margins are seen (Figure 335). An island of bone, completely detached and somewhat denser than normal, indicates a sequestrum (Figure 336). It is often impossible to differentiate osteomyelitis from eosinophilic granuloma or other tumors of the skull by x-ray examination alone.

Prior to the advent of chemotherapy the only satisfactory treatment of osteomyelitis of the skull consisted in radical excision of bone well beyond the grossly involved area, sometimes necessitating very extensive craniectomy (Figure 337). Established infection, particularly in bone whose blood supply is at all impaired, as in an infected bone flap or post-traumatic



Figure 335 Lateral roentgenogram of 10 6/12 year old child showing extensive osteomyelitis of the frontal and parietal bones secondary to acute sinusitis.



Figure 336. Lateral roentgenogram of the skull after coronal craniectomy for premature fusion of the coronal suture. Arrow points to sequestrum associated with a chronic draining wound sinus. Successfully treated by removal of sequestrum and simultaneous chemotherapy.



Figure 337. Lateral roentgenogram of the same patient shown in Figure 335 following extensive craniectomy for complete removal of infected bone.

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Chapter II

Brain Abscess

BRAIN ABSCESS in childhood results usually from one of three sources: (1) direct or venous extension from pre-existing infection in the paranasal air sinuses, middle ear, or mastoid; (2) hematogenous metastasis from pre-existing pyogenic infection elsewhere in the body, principally in the lungs and pleura; and (3) compound fractures of the skull with contaminated foreign bodies or bone fragments driven into the cerebral substance. Two other varieties of brain abscess which occur in children deserve particular mention. The first of these is abscess associated with an infected dermoid cyst, usually in the cerebellum. This infection is due to direct extension from the skin by way of a persistent congenital dermal sinus. These have been described elsewhere (p. 69). The second special type of abscess in childhood has actually been the most common variety seen in this clinic in the last two or three years; that is, the so-called "paradoxical" cerebral abscess associated with congenital cyanotic heart disease. This will be discussed separately in a subsequent section.

Like osteomyelitis of the skull, pyogenic abscess of the brain has markedly diminished in frequency since the advent of modern chemotherapy. While 93 patients have been treated for brain abscess in this hospital, there have been only two or three per year since 1949. The immediate cause for this reduction, of course, is the vastly improved treatment of otitis, mastoiditis, sinusitis, lung abscess, empyema, bronchiectasis, and pyogenic meningitis. It is important to remember, however, that brain abscess secondary to these or any other distant sources of infection does still occur occasionally and that, with the widespread use of antibiotics, the clinical features of the lesion may be masked and go long unrecognized. It is necessary, therefore, to keep abscess in mind in the differential diagnosis of increased intracranial pressure or focal neurological abnormalities, even if fever, chills, leucocytosis and other systemic evidences of infection are lacking. Because of the infrequency of this type of brain abscess in children now and because the management differs little from that appropriate to adult practice, the problem will be presented here only briefly.

fragment, is still treated best by complete removal of the involved bone. With antibiotic support, however, one is justified in performing a much more limited craniectomy. When x-ray studies indicate the presence of a sequestrum associated with a chronically draining wound sinus, surgical removal is always indicated. Early and continued use of proper chemotherapy should obviate the necessity for surgical excision of any but previously devitalized bone in most instances.

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Clinical Features

A clinical history or other evidence of infection is usually present but may be so minimal that it is not volunteered by the child or his parents. Frequently an antecedent infection may have been disregarded because it had apparently been brought quickly under control by chemotherapy. Inquiry should therefore be made specifically for symptoms and signs suggestive of otitis, sinusitis, mastoiditis or pulmonary infection. Rarely nowadays is there a definite phase in the course of brain abscess development indicative of a bacteremia, and there may be a long latent period between initial infection and the appearance of cerebral involvement.

The symptoms and signs of brain abscess may be divided into three groups; those relative to infection, those pertaining to increased intracranial pressure and those associated with focal neurological disturbance. As already emphasized, in many children the symptoms and signs may be extraordinarily few.

The signs due to infection during the acute phase of intracranial suppuration may include fever, malaise, poor appetite, leucocytosis, stupor and meningismus. In a chronic, well-encapsulated brain abscess, there may be no clinical indication of infection.

Symptoms and signs of increased intracranial pressure are perhaps the *most consistent. Headache, vomiting, irritability, drowsiness, papilledema,* and separation of the sutures in infants may all be present. Increased pressure may become extreme with unusual rapidity in this lesion leading to profound coma, respiratory and cardiac irregularity and death if untreated.

Focal neurological signs depend obviously on the location of the abscess. Lateralizing convulsive seizures may occur. Cerebral abscess secondary to ear and mastoid infection is most commonly located in the temporal lobe. In this location there may be no abnormal neurological signs. Homonymous visual field defects are extremely difficult to determine in infants and young children. Aphasia and hemiparesis are perhaps the most reliable focal signs. Cerebellar abscess is usually accompanied by hypotonia, nystagmus and ataxia in addition to evidence of increased intracranial pressure. Electroencephalography may give a clue to localization occasionally in the absence of clinical signs.

Plain x-ray studies are significant in indicating increased intracranial pressure and also in helping localize a possible source of infection. Thus, films of the sinuses, mastoids and chest are of importance in addition to skull films when brain abscess is suspected. Ventriculography should be employed promptly for diagnosis and localization when this is not possible by clinical means.

Spinal fluid findings on lumbar puncture vary considerably but are seldom normal. In an acute brain abscess there are usually many white blood cells (100 to 1,000 per cu. mm.) with a high percentage of polymorphonuclear cells. In a chronic encapsulated abscess, there may be very few abnormal cells in the spinal fluid and these are usually lymphocytes (10 to 100 per cu. mm.). The total protein content is almost invariably elevated moderately (50 to 150 mgm. per cent). Spinal fluid cultures are ordinarily negative and the sugar may be normal. The pressure may be exceedingly high and is rarely apt to be normal. Removal of spinal fluid in the presence of increased pressure due to a cerebellar abscess may be extremely hazardous and should be avoided.

Treatment

The treatment of solitary brain abscess after proper localization is nowadays much more satisfactory and direct than formerly. Three methods of therapy are used in this clinic depending on the individual characteristics of the abscess. These are: (1) repeated tapping through a burr hole, (2) continuous catheter drainage after initial aspiration through a burr hole, and (3) total excision of the abscess. It should be emphasized that intensive antibiotic therapy with appropriate drugs depending upon the bacteriology of the abscess and the original source of infection must be pursued throughout whichever method of surgical treatment is employed. It should also be emphasized that newer methods of treatment of brain abscess have not altered the fundamental surgical principle of initial adequate treatment of the focus from which the brain abscess developed.

Repeated Tapping: If an abscess is located far from the surface and particularly if there is not a well-defined tough capsule, this may be the treatment of choice. When the abscess is first tapped, as much purulent material as possible is removed, cultures are made, and the cavity is irrigated gently with saline or penicillin solution. Before removal of the aspirating cannula, 2 or 3 ccs. of colloidal sodium thorotrast are injected. It may be helpful to inject 5 to 10 ccs. of air at the same time. Roentgenograms made immediately with the head in various positions then outline the position of the abscess very accurately (Figure 338). Since thorotrast is picked up by phagocytic cells in the abscess wall, subsequent films may be made to follow the lesion. The abscess is tapped at intervals determined by the patient's course, the roentgenographic appearance and the amount of pus recovered. Taps are continued until nothing more can be obtained and the cavity remains collapsed by x-ray examination.

Catheter Drainage: After initial aspiration a rubber catheter, usually size No. 12 to No. 16 French, is introduced into the abscess lumen. It is wise to insert the catheter before attempting to empty the abscess cavity in

order to facilitate its proper placement. The catheter should be loosely secured to the skin surface to prevent its coming out; it should never be fastened tightly to the dura because of the danger of its tip being pushed through the distal wall of the abscess into uninfected brain or into the ventricular system. A bulky dressing is used and the catheter is irrigated one or more times daily to prevent clogging. Thorotrast may also be used with this method to follow the size and position of the abscess lumen by x-ray (Figure 339). Catheter drainage is particularly suited to cerebellar abscesses and to superficial cerebral abscesses which are poorly encapsulated.

Total Excision: The most desirable method of treatment of brain abscess is total excision by open craniotomy, thus completely eradicating the area of infection. With antibiotic therapy, it is possible to bring a much higher proportion of brain abscesses to a chronic well encapsulated stage where this type of treatment is possible. Tapping or catheter drainage as outlined, supported by antibiotic protection, may sometimes carry a child through the precarious phase of increased intracranial pressure and severe toxemia and permit subsequent radical excision when there has been an improvement in general condition. It is always wise to visualize the abscess by x-ray after thorotrast injection if possible before performing craniotomy. Radical excision is carried out in the same manner as removal of any benign



Figure 338. Antero-posterior and lateral roentgenogram of 4 8/12 year old boy after aspiration of a large mid-line subfrontal abscess associated with ethmoiditis. Five ccs. of thorotrast and 5 ccs of air were injected into the abscess cavity after removal of 65 ccs. of thick purulent material. Shrinking of the abscess wall was followed by subsequent x-ray studies

cerebral tumor. The lesion is approached by the shortest available route through silent cortex. When the abscess capsule is defined, careful dissection is continued in the thin layer of necrotic brain just outside it until the entire abscess is delivered intact. If the wall is tough, this may be quite simple; if it is thin, great care must be taken to retract the brain gently and tip the abscess out without rupture (Figure 340). After such excision, the dura is closed tightly and the rest of the wound approximated without drainage. Full antibiotic and anticonvulsant protection should be maintained throughout the operative and post-operative periods.

The prognosis for single abscess of the brain with modern methods of treatment is vastly improved if the correct diagnosis is made before the abscess ruptures into the ventricular system or subarachnoid space and before irreversible brain stem changes due to severe increased intracranial pressure have occurred. With continued improvement in the chemical treatment of pyogenic infections, brain abscess may well soon become a surgical curiosity.

Brain Abscess in Congenital Heart Disease

One variety of brain abscess of special significance in childhood is that associated with congenital heart disease. This form of metastatic abscess is always associated with a cardiac defect which permits venous blood to

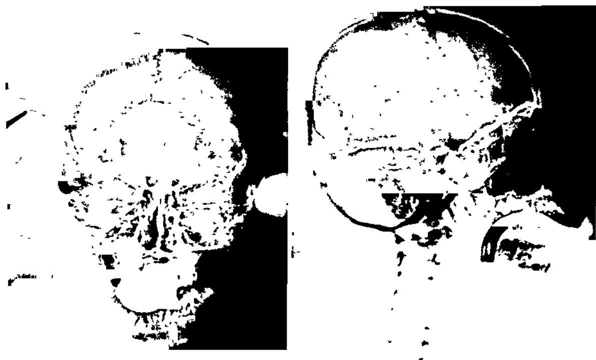


Figure 339 Antero-posterior and lateral ventriculogram made one month after drainage of right posterior temporal lobe abscess. The abscess capsule and tract of drainage catheter are outlined by thorotrast. Deformity of the right temporal horn of the ventricular system is well shown.



Figure 340. Encapsulated brain abscess removed intact from the temporal lobe together with sinus tract which had formed along the path of previous aspiration This abscess was secondary to mastoid infection in a nine year old child.

circulate in the arterial system without passing through the lungs, that is, with so-called right-to-left shunt, or cyanotic heart disease. In these patients there is polycythemia, with red blood cell counts of 5.0 to 7.5 million and hemoglobin values of 15.5 grams to as much as 20 grams, usually some clubbing of the fingers, transient or permanent cyanosis, and very often small stature and poor exercise tolerance. By far the commonest variety of cardiac anomaly associated with these paradoxical cerebral abscesses has been Tetralogy of Fallot (Figure 341). It has also been reported with other types of interventricular defect, persistent truncus arteriosus and cor triloculare.

Although the association of cerebral abscess with congenital heart disease was first recorded in 1814 [67] and has been frequently noted since then in pathological studies, it is only in recent years that the condition has been diagnosed before death and successfully treated surgically.[32, 83, 102] Gluck, Hall and Stevenson [73] have recently reviewed the available literature and found 44 indisputable cases, of which 20 were under 10 years of age. In 90 per cent of all the cases the abscess was single. In their collected cases, there were only 10 treated by cranial operations and only four of these were successful.

In this clinic six patients have been seen with brain abscess and cyanotic heart disease since 1948. In the first two, the diagnosis was made at au-

topsy. In the third, an emergency drill hole was made and the abscess tapped as soon as the patient was seen in consultation, but the child already had dilated fixed pupils and had been artificially respirated for some time; spontaneous respirations were never resumed. The fourth patient was transferred from another hospital moribund with severe meningitis and extreme papilledema. The abscess was tapped and subsequently excised, but after a lingering course, the patient finally succumbed (Figure 342). In the fifth and sixth patients, the correct diagnosis was made fairly promptly after the onset of signs of increased intracranial pressure and focal neurological disturbance and successfully treated by drainage and excision supported by antibiotic therapy (Figures 343 and 344). One of these patients subsequently was operated upon for correction of the Tetralogy of Fallot with success (Figure 345).

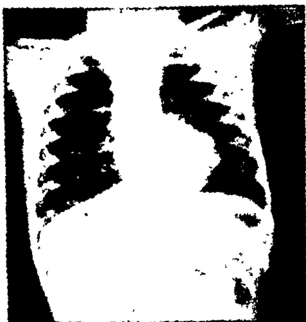


Figure 341. Chest roentgenogram of 2 1/12 years old child with Tetralogy of Fallot who had a left temporal lobe brain abscess. Note right ventricular hypertrophy, right aortic arch, deviation of trachea.

Etiology

It is generally accepted that infection reaches the brain from a distant focus either as paradoxical infected emboli or as secondary infection of a previously infarcted area of brain. It is probable that decreased blood flow due to increased viscosity secondary to polycythemia and also decreased pulmonary blood flow secondary to the congenital cardiac defects themselves favor the formation of thrombi. The source of infection is problematical. Among Gluck et al.'s reviewed cases, only eight had a definite history of recent infection, in five there was a vague history of upper respiratory infection and in the other 30 there was no suggestion of infection elsewhere. Blood cultures have been negative whenever reported; they were negative in our cases. In many of these patients, cultures of the abscess itself have been negative. The commonest organism reported is an anaerobic *streptococcus*. It is of interest that in vitro this is an organism which grows well in brain broth. Perhaps there is often transient bacteremia due to these organisms which in normal individuals is ordinarily overcome readily in the capillary bed of the pulmonary circulation. When the latter is by-passed through a right-to-left shunt the organisms reach the brain and are able to survive in this tissue in polycythemic patients.



Figure 342. Large parieto-temporal abscess as seen by ventriculography after previous injection of thorotrast into the abscess at the time of aspiration. This occurred in a 2 7/12 year old infant with Tetralogy of Fallot.



Figure 343 Antero-posterior and lateral roentgenograms of the skull of an 11 8/12 year old girl with occipital lobe brain abscess associated with cyanotic heart disease. Air and thorotrast have been injected into the abscess lumen after aspiration of 60 ccs. of purulent material. Abscess well localized by this means and subsequently excised at craniotomy (Figure 344).



Figure 344. Large encapsulated brain abscess removed intact from the occipital lobe of an 11 8/12 year old girl with cyanotic heart disease due to persistent truncus arteriosus Patient had increased intracranial pressure and homonymous hemianopsia.

Clinical Features

In every patient with cyanotic congenital heart disease, the development of convulsions, focal neurological symptoms and signs, or evidence of increased intracranial pressure, the possibility of cerebral abscess formation should be considered. As already pointed out, there may be no history of infection and there is usually no fever, leucocytosis, or clinical evidence of systemic infection. The commoner symptoms are headache, vomiting and lethargy. The development of focal neurological signs such as hemiplegia, aphasia, or hemianopsia is particularly significant and always demands investigation. There may or may not be evidence of mild meningeal irritation. Lumbar puncture has always been abnormal in our experience showing increased pressure and increased numbers of white blood cells, but negative culture. Roentgenograms of the skull are normal. Electroencephalograms should be localizing if a large abscess is present.

Treatment

Since almost all of these abscesses have been single, surgical treatment supported by antibiotic therapy should be successful in a high percentage of cases if the diagnosis is made before rupture or irreversible cerebral damage has occurred. If localization cannot be made on the basis of clin-

ical and electroencephalographic findings, ventricular air studies should be performed promptly. A burr hole should then be placed over the lesion and needle aspiration carried out. If the abscess is encapsulated, subsequent surgical excision is recommended. Injection of 2 or 3 ccs. of colloidal sodium thorotrast into the abscess cavity at the time of initial drainage serves to outline the limits of the abscess on x-ray examination and facilitate placement of a bone flap for surgical removal (Figures 343 and 344).



Figure 345. Three and eleven-twelfths years old girl, 19 months after drainage of temporal lobe brain abscess and 13 months after surgical treatment for Tetralogy of Fallot (Pott's procedure).

Chapter III

Subdural Effusion Complicating Meningitis

N 1948 IT FIRST became evident that accumulation of high protein xanthochromic fluid in the subdural space, often becoming encapsulated, is a frequent complication of bacterial meningitis.[140] Since that time, in this and other clinics, increasing numbers of infants with this complication have been discovered.[5, 6, 19, 139, 182, 185] In our experience subdural effusion has most commonly followed meningitis due to *h. influenzae*. It has also been a sequel occasionally, however, to *meningococcus*, *streptococus* and *pneumococcus* meningitis (Table XI).

TABLE XI
TYPES OF MENINGITIS ASSOCIATED WITH SUBDURAL EFFUSION

	No. of Cases
H. Influenzae	25
Pneumococcus	10
Meningococcus	7
Staphylococcus	1
Tuberculous	1
No Organisms Identified	6
Total	50

The cause of this subdural fluid accumulation is not clear. Its incidence is greatest during the first year of life (Figure 346), but it has been found in older children as well. It is probably seen with increased frequency recently because the incidence of infants recovering from the acute phase of severe bacterial meningitis is much greater since the advent of modern antibiotic therapy.

It seems probable that there is an exudation of fluid into the subdural space in response to the irritation of infection in the adjacent pia-arachnoid. Since this fluid is trapped within a space from which there is little absorption, the protein content becomes very high and an isolating membrane tends to form around it as it does around blood in the subdural space (p. 191). Whether antibiotic therapy itself or simply repeated lumbar punctures in treatment of the meningitis play any role in the etiology of

these fluid collections is unknown. It has occurred following various types and combinations of antibiotic treatment. There is frequently some evidence of recent bleeding into the fluid, perhaps from congested vessels in the infected meninges or from bridging veins stretched by enlargement of the normally potential subdural space.

The possibility of subdural effusion complicating meningitis, particularly that due to *h. influenzae*, should be suspected in any infant not responding quickly and satisfactorily to adequate antibiotic treatment. There has been no uniform clinical picture. The commonest finding is persistent fever with general irritability and failure to eat well and gain weight after apparent bacteriologic cure. The diagnosis is made as in traumatic subdural hematoma by bilateral subdural puncture through the coronal suture. In almost every instance the subdural effusion has been bilateral. It is our feeling that such exploratory taps should be performed on any infant with meningitis whose clinical course is unsatisfactory generally or who shows any of the following: (1) fever after 48 to 72 hours of adequate specific and supportive treatment; (2) persistent positive spinal fluid cultures; (3) focal or persistent convulsions; (4) persistent vomiting; or (5) specific neurological abnormalities after subsidence of infection.

SUBDURAL EFFUSION IN INFANTS WITH MENINGITIS

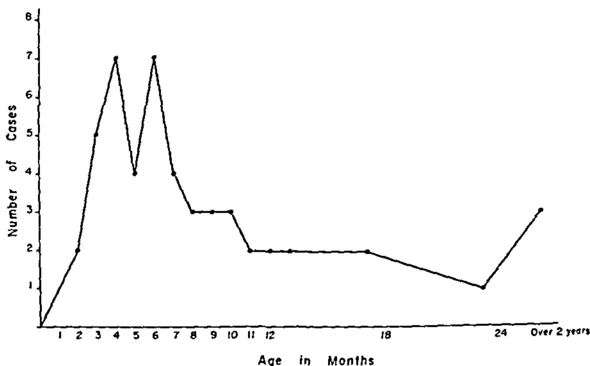


Figure 318 Age distribution of 50 infants who developed subdural effusion of high protein, xanthochromic fluid associated with meningitis

Treatment of this complication is essentially the same as that outlined for traumatic subdural hematoma (p. 196). The first diagnostic subdural tap begins treatment, and the infant may start to improve immediately. If started early, repeated subdural taps will usually dry up the effusion quickly and no further treatment is indicated. In more chronic accumulations, the fluid may be deeply yellow, contain some blood and have a very high protein content. Under these circumstances it is usually encapsulated between an outer and an inner membrane. In early infancy it is necessary to remove these membranes by craniotomy to prevent reaccumulation of the fluid and to permit normal re-expansion and growth of the brain.

In all patients, therefore, who repeatedly demonstrate large amounts of yellow subdural fluid after meningitis, a craniotomy is planned under general anesthesia. The incision is outlined and the superior frontal burr hole of the proposed bone flap is drilled. If this demonstrates that a membrane is present, the craniotomy is carried out; if there is no membrane, the subdural space is irrigated thoroughly through one or more burr holes and the same procedure then repeated on the other side at the same operation. If craniotomy is performed, the second side is explored in seven to 10 days.

It is wise to delay surgery and continue subdural taps until the patient is fever-free, cultures of the subdural fluid and lumbar cerebrospinal fluid are negative and the cell counts of these fluids are reduced to below 100 w.b.c./cu.mm. Antibiotic therapy should be continued through the period of subdural tapping and operative treatment.

At operation the membrane is apt to be much more ragged and adherent than that seen in subdural hematoma. Collections of fibrin in the subdural and subarachnoid spaces are often seen. After operation these patients should be kept on anticonvulsant therapy for at least the first two weeks even if there have been no pre-operative seizures. If not, a high percentage have had one or more generalized seizures in our experience. Otherwise the general plan of operative and post-operative treatment is that described elsewhere for subdural hematoma (p. 196).

Our surgical experience with this complication now includes over 50 patients. The type of meningitis is shown in Table XI. In this group of patients, 15 were treated by subdural taps alone and 35 in addition by craniotomy with removal of subdural membranes. Fourteen patients had subdural membranes bilaterally. The immediate results on the whole have been very gratifying. Unless there was severe brain damage at the time of operation, the patient's general condition as well as his neurological abnormalities usually responded promptly and dramatically. Long term follow-ups are not as yet available, but there is every reason so far to be encouraged with the progress of these infants.

PART VII

CEREBRO-VASCULAR DISORDERS

Chapter I

Angiography

The indications and techniques for angiography in children vary little from those widely discussed in recent years in adult neurosurgical practice. The procedure is safe in infancy and childhood and in our clinic has been performed in all age groups from a few weeks up without serious complication (Figure 347).

Obviously in infants and in most children less than 12 years of age, it must be performed under general anesthesia. In older children it has usually been possible to use a percutaneous injection technique as generally practiced in adults, but in infants and younger children we have uniformly resorted to direct injection after surgical exposure of the common carotid artery through a small incision in the line of the crease of the neck (Figure 348). The simplest possible draping technique is recommended to facilitate satisfactory positioning of the child's head during exposure of the films (Figure 349).

It is our practice to use 35 per cent diodrast, injecting from 4 ccs. to 10 ccs. of solution for each series of pictures. We have used an automatic seriograph when it was desirable to study both arterial and venous phases of circulation (Figure 350) and carried out separate injections for anteroposterior and lateral views. The injection is made as rapidly as possible through a No. 18 or No. 19 gauge needle with the full, unoccluded stream of carotid blood flow. If the bevel of the needle is directed posteriorly and laterally maximum flow into the internal carotid artery will occur following common carotid injection. It is not necessary to perform the more difficult exposure of the internal carotid artery itself. Vertebral arteriography in children has always been performed directly in this clinic after operative exposure of the vertebral artery through an incision similar to that used for carotid exposure (Figure 348). We have not used sympathetic blocks, papaverine administration, or other supportive measures at the time of injection in children. In angiography in more than 100 children under 12 years of age we have not experienced convulsions, hemiplegia or fatality. Occasionally bilateral injections have been performed under the same anesthesia and as much as 40 to 50 ccs. of 35 per cent diodrast have been injected into small children without difficulty. We have also occasionally

Figure 347. Antero-posterior and lateral views of a normal carotid arteriogram in a six months old infant.

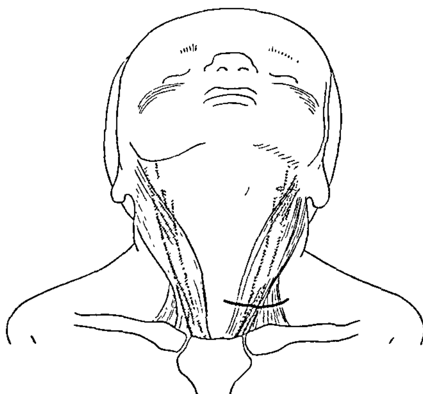


Figure 348. Location of incision to expose the common carotid artery for cerebral angiography in infants and young children The sternomastoid muscle is retracted laterally.

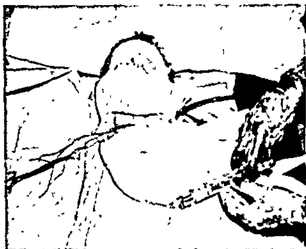


Figure 349. Carotid arteriography using 10 ccs. of 35 per cent sodium diodrast. Note method of draping for surgical exposure of carotid artery using small plastic drape so that head is well exposed for correct positioning on x-ray table. Separate injections are made for the antero-posterior and lateral projections. Rubber tube may be eliminated for more rapid direct injection.



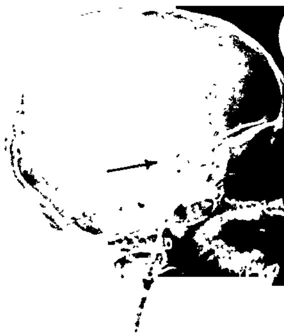


Figure 350 (*Left*). Late venous phase of carotid arteriogram of six months old infant.

Figure 351 (*Right*). Lateral arteriogram after injection of the left carotid artery in a 3 4/12 years old child with right hemiplegia of three weeks' duration. Note narrowing of the arteries at the region of the bifurcation and failure of the main branches of the middle cerebral artery to fill. This pattern was consistent in three separate injections. Patient's subsequent history and gradual recovery were consistent with the diagnosis of thrombosis of the middle cerebral artery.

performed air studies and arteriography under the same anesthesia without mishap.

Angiography has been employed with increasing frequency for the diagnosis and study of arterial aneurysms (Figure 352), arteriovenous malformations (Figure 354), cerebral tumors (Figure 248) and convulsions and hemiplegias of unknown etiology (Figure 351).

Chapter II

Congenital Malformations

ARTERIAL ANEURYSMS

Arterial aneurysms of the Circle of Willis have rarely been demonstrated as a cause of focal neurological signs or subarachnoid hemorrhage in infancy and early childhood. Although it is believed that these sacular or "berry" aneurysms occurring at or near points of bifurcation of the major arteries at the base of brain are of congenital origin, they do not often become symptomatic before 12 years of age. Presumably they do not grow to sufficient size to rupture until at least the latter part of the second decade.

In our experience, spontaneous subarachnoid and intracortical hemorrhage have occurred much more frequently in childhood from cortical arteriovenous malformations or else from entirely undetermined sources.



Figure 352. Lateral carotid arteriogram in 7 10/12 year old boy showing huge sacular aneurysm apparently arising at the bifurcation of the anterior and middle cerebral arteries. Arteriogram made 10 days following severe subarachnoid hemorrhage. Patient was treated by ligation of the internal carotid artery and has been asymptomatic without further bleeding in the year since operation.

It is possible that in early life rupture with fatal hemorrhage may occur from an aneurysm so small that it is not demonstrated by arteriography or even by meticulous post-mortem dissection. Only one sacular aneurysm of the Circle of Willis has been demonstrated by arteriography in this clinic in a child under 12 years of age. This was in a boy of seven who had a massive subarachnoid hemorrhage resulting in profound coma, mild left hemiparesis and transient right third nerve palsy. There was a moderately loud systolic bruit heard over the right eye and over the right anterior temporo-frontal region. Right carotid arteriogram revealed a large aneurysm arising near the bifurcation of the anterior and middle cerebral arteries (Figure 352).



Figure 353. TBE x 100 Photomicrograph of section through arterio-venous malformation removed from the left parietal cortex of a 12 year old boy. This is an irregular mass of blood vessels, some of which have thick arterial walls with elastic and muscular layers, while others have only a thin membrane and endothelial lining surrounded by a connective tissue sheath. There is some lymphocytic infiltration between the blood vessels.



Figure 354. Antero-posterior and lateral roentgenograms following carotid injection of diodrast showing arteriovenous malformation in the left parietal cortex of a 12 year old boy.

In spite of its rarity, arterial aneurysm must always be considered as a possible source of spontaneous subarachnoid hemorrhage in childhood. In the absence of localizing signs on neurological examination, bilateral carotid arteriography should be performed.

ARTERIOVENOUS MALFORMATIONS

Clinical Features

Arteriovenous malformations of the cerebral hemispheres occasionally become symptomatic in early childhood. They consist usually of a tangled collection of blood vessels of varying size (Figure 353) either diffusely distributed over the surface of the cortex or grouped into a more discrete mass within the confines of the brain.[47] These vessels are in the pia-arachnoid for the most part so that they are not actually in the substance of the brain but instead are intimately applied to the surface of its convolutions. Often, the larger feeding arteries and draining veins course on the exposed outer cortex, and the main mass of the anomaly lies within the cerebral hemisphere or in one of the interlobar fissures (Figures 354 and 355).

Another variety of arteriovenous malformation that has been reported in children a number of times is that involving the posterior cerebral ar-





Figure 355. Arteriovenous malformation.

A. Carotid arteriogram in a 6 6/12 year old girl showing abnormal connections from external carotid and posterior cerebral arteries.

B. Vertebral arteriogram in [illegible] ent showing huge malformation in occipital region [illegible] branches of the basilar and posterior [illegible] s.

teries and the vein of Galen. This is characterized by a large aneurysmal dilatation of the artery which because of its location has led to obstruction of the Aqueduct of Sylvius with internal hydrocephalus. Of those reported, some have been fed by one artery only and some by connections from both sides. Drainage has been usually directly into a much enlarged vein of Galen or straight sinus (Figure 356).

These lesions are undoubtedly *congenital abnormalities* in the development of the embryonic intracranial blood spaces. To what extent further increase in relative size or complexity of the malformation occurs after birth in unknown. They may become symptomatic at any age.



Figure 356. *Post-mortem specimen of large arteriovenous aneurysm between the posterior cerebral artery and the vein of Galen in a 20 months old infant who had extreme hydrocephalus.*

Cerebral arteriovenous malformations make their presence known by a variety of symptoms and signs, including: (1) headache; (2) focal neurological abnormalities; (3) convulsive seizures; (4) spontaneous subarachnoid or intracortical hemorrhage; and (5) subjective or objective detection of an intracranial bruit.

In infants and children, *headache*, vomiting and other evidence of increased intracranial pressure are not usually a feature of cerebral vascular anomalies because there is no sudden increase in mass and ordinarily no obstruction to the cerebrospinal fluid pathway. As already noted such obstruction may occur rarely with vascular lesions about the brain stem and in the posterior fossa.

Focal neurological signs such as hemiparesis, aphasia, or homonymous visual field defects may result from large cortical vascular malformations. Certainly the presence of these signs in early life in the absence of increased intracranial pressure, trauma or infection should arouse suspicion of this possibility.

One of the commoner symptoms leading to detection of these lesions is the occurrence of *convulsive seizures*, particularly when they are consistently focal in character. Repeated focal seizures in a child without intracranial pressure, high fever, history of trauma or other apparent ab-

normality should always suggest a cortical vascular anomaly. The electroencephalogram, if positive, may show a well localized spike, or spike-and-wave disturbance. Presumably, disturbance of blood supply to or local compression of an area of cerebral cortex leads to gliosis and neuronal changes that constitute an epileptogenic focus (Figure 357).

Perhaps the commonest and certainly the most serious complication of these malformations in childhood is *spontaneous intracranial hemorrhage,* either intracortical, subarachnoid, or both. Indeed, the first indication of any abnormality may be sudden occurrence of severe pain in the head or neck followed by screaming, vomiting and rapid appearance of hemiplegia or coma. What causes these malformations to bleed at any one particular moment is pure conjecture. Certainly in childhood they are not associated with arterial hypertension and the hemorrhage does not necessarily occur at any time of unusual physical or emotional exertion. If the hemorrhage is entirely into the subarachnoid space, there is apt to be complete absence of localizing neurological signs. There is headache, stiffness of the neck, hyperactive reflexes and in severe cases, profound loss of consciousness and interference with vital functions. If the hemorrhage is intracortical, hemiplegia, aphasia, hemianopsia or convulsions are apt to ensue. Initial fatal hemorrhage is much less apt to occur than from a saccular aneurysm of the Circle of Willis. Several instances of repeated hemorrhage from an arteriovenous malformation have occurred in our clinic.

An intracranial *bruit* may sometimes be heard in infants in the absence

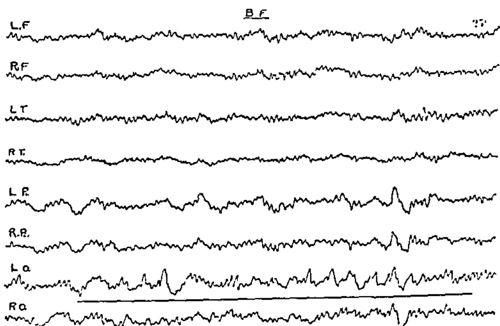


Figure 357. Electroencephalogram of 13 year old boy with subcortical arteriovenous malformation in the left parieto-occipital region.

of a vascular malformation. This is particularly true in the presence of increased intracranial pressure where splitting of the sutures makes increased blood flow to the head audible. Ordinarily, though, an audible bruit, especially if it is unilateral, indicates a blood vessel anomaly. Not all arteriovenous cortical malformations are accompanied by an audible bruit by any means, and its absence never rules out this anomaly. However, careful auscultation over the entire cranial vault including the orbits should be performed in all children who are suspected of central nervous system pathology.

The diagnosis of arteriovenous malformation may be suspected on the basis of history, examination and the occurrence of hemorrhage, but it is confirmed definitely only by cerebral arteriography or surgical exploration. Plain roentgenograms of the skull are usually negative. Carotid or vertebral arteriography is performed as already described (p. 395) and usually demonstrates the extent and nature of the anomaly (Figures 354 and 355). Serial films in different projections, to demonstrate the location of the principal feeding arteries, are exceptionally valuable in this lesion if surgical treatment is to be undertaken.

Treatment

Until recent years surgical treatment of cerebral arteriovenous malformations was rarely attempted. However, with improved surgical methods of hemostasis, induced hypotension, availability of large amounts of blood for rapid infusion and particularly with careful pre-operative arteriograms to demonstrate the extent and position of the arterial supply of the lesion, an increasing proportion of them should be surgically treatable. Since these anomalies are located well beyond the major arterial bifurcations at the base of the brain, ligation of the carotid artery in the neck is probably of no avail. Other methods of attack have been: (1) to shrink the size of the major vessels gradually by stroking them with low-voltage electrocautery,[187] (2) to expose the major feeding arteries intracranially and simply ligate these, (3) to perform a lobe or wedge resection of the cerebrum including the area of the vascular anomaly after securing the principal blood supply,[161] and (4) subpial, meticulous dissection and excision of all the abnormal vessels, ligating and dividing the feeding arteries first and the draining veins last without removal of any cerebral tissue.[143] The last is the method of choice whenever possible (Figures 358 and 359). Although the mass may shrink considerably after ligation of a single large feeding artery, there are usually several arterial connections, and it is not until the last of these is divided that the arteriovenous shunt is actually obliterated.



Figure 358 (*Upper*) Arteriovenous malformation of the cortex exposed at operation. Subpial dissection of the malformation is almost complete. The shrunken lesion can be seen in the depths of the operative exposure. Note, however, that the principal draining vein to the longitudinal sinus at the bottom of the picture still contains arterial blood.

Figure 359 (*See legend on facing page.*) →



Figure 360. Lateral carotid arteriogram showing displacement of the middle cerebral vessels associated with large spontaneous hemorrhage into the left temporo-parietal area in an 11 6/12 year old girl. No source for the hemorrhage was ever demonstrated. Satisfactory recovery following evacuation of huge intracortical hematoma.

SPONTANEOUS INTRACEREBRAL HEMORRHAGE

An additional note is presented here regarding the occurrence of spontaneous intracerebral hemorrhage of unknown etiology. Our experience with this baffling situation in childhood has been disturbing. Perhaps the role of small subcortical congenital vascular malformations is greater than has been suspected.[130]

In this clinic we have seen in recent years at least eight to 10 children under 12 years of age with massive hemorrhage into the white matter of one cerebral hemisphere in whom successful evacuation of a huge hematoma has been performed without ever demonstrating either at operation or by subsequent arteriography any tumor or vascular abnormality. Most of these large clots have been found in the posterior temporo-parietal area (Figure 360). There has been no history or clinical evidence of trauma. The onset has been sudden and the symptoms of increased intracranial pressure and hemiplegia usually rapidly progressive.

←

Figure 359 (*Lower*) Same patient as in Figure 358. The last arterial connections to the arteriovenous malformation have been divided and the lesion removed. Note now the venous character of the blood in the principal vein draining the area Note also that no brain has been excised in the removal of this lesion.

There has seldom been indication or opportunity for pre-operative arteriography or ventriculography in these patients and emergency evacuation of the clot has in some been a life-saving procedure. In none of these children in whom a large hematoma was successfully evacuated and no source for the bleeding identified has a second hemorrhage occurred to our knowledge. When this lesion is present, aspiration of subcortical clot through a brain cannula is effective only for diagnosis and localization. Adequate evacuation should then be carried out by direct transcortical exposure of the clot through the most silent area of brain which is available.



Figure 361. Persistent parietal foramen with emissary communications between the longitudinal sinus and dilated scalp veins.

PERSISTENT EMISSARY VEINS

Ordinarily, as embryonic growth proceeds, the free communication between extracranial and intracranial venous circulation present in the young fetus is obliterated except for small parietal and mastoid emissary veins. Occasionally one of these connecting veins in the postparietal or occipital area remains unusually large. Under these circumstances, whenever intracranial pressure is increased, there is a reversal of flow so that blood from intracranial venous sinuses enters the scalp veins, and the latter become dilated and tortuous (Figure 361).

At rest, particularly when the head is elevated, these scalp veins are collapsed and hardly apparent. When the child cries or strains, however, the veins may become alarmingly congested (Figure 362). The same effect can be produced by bilateral jugular compression.

Before interrupting these abnormal emissary vessels, it has been our custom to inject 35 per cent diodrast into the scalp veins in order to demonstrate that normal intracranial venous sinuses with drainage into the internal jugular veins exist (Figure 363). It is then a simple matter to expose the persistent emissary foramen by a curved scalp incision. The connecting scalp veins are dissected free and divided between ligatures at the level of penetration through the skull defect. A flap of periosteum

from adjacent bone is elevated and inverted over the skull defect to reinforce the closure. No post-operative complications have been experienced.



Figure 362. Six months old infant with persistent postparietal foramen and huge emissary venous communication between the intracranial and extracranial circulation. Note the marked dilatation of connecting veins when the baby cries. Satisfactorily treated by interruption of the communication at the level of the dura.



Figure 363 Lateral roentgenogram during diodrast injection into the scalp veins of six months old infant showing its passage into the longitudinal sinus through persistent emissary connection Both transverse sinuses and the internal jugular veins are well visualized.

Chapter III

Sturge-Weber Syndrome

In 1879, Sturge[191] reported the case of a six and a half year old girl who was born with a "port wine" stain on the right side of her face and scalp who began to have contralateral seizures at the age of six months. He postulated: "From the nature of the fits, and from their mode of onset, I think there can be no doubt that they are due to some cause external to the nerve tissue, rather than to an inherent irritability of grey matter, and this external cause in all probability to be found in the presence of a 'port wine mark' on the surface of the right side of the brain, just as we have found it in the skin, mucous membranes, and retina of that side." In 1922 and again in 1929, Weber[201] described a patient with this syndrome whose skull was small on the side of the cutaneous angioma and who showed by x-ray examination extensive calcification in the cerebral hemisphere of this side. Since that time the literature has contained numerous reports of examples of this "neurocutaneous syndrome"[31, 40, 75, 77, 113, 206]

The lesion is presumably due to, or associated with, a congenital ectodermal defect since it seems to involve corresponding segments of the skin and central nervous system. The cutaneous lesion is evident at birth but involvement of the central nervous system may not manifest itself until much later.

The essential clinical features of the syndrome include the following:

Cutaneous Nevus: This is commonly a cavernous rather than telangiectatic type of cutaneous angioma which varies from port-wine to deep purple in color. It is usually flat and sometimes quite pale and barely discernible. It most commonly involves the face, particularly the area of the first and second divisions of the fifth cranial nerve, seldom crossing the mid-line (Figure 364). It may involve the neck, trunk and extremities much less frequently (Figure 365).

Convulsive Seizures: These may be either general or focal, involving the side of the body opposite the cutaneous lesion. In young children they are frequently of the petit mal variety. Often the seizures increase in frequency and severity as the child grows and occasionally they may be extremely difficult to control with anticonvulsant medication.

Hemiplegia: This usually takes the form of a spastic hemiparesis contralateral to the side of the cutaneous lesion. As growth proceeds, there may be hemi-atrophy or retarded growth of the hemiparetic side of the body. In older children, homonymous hemianopsia is also frequently detectable.

Mental Retardation: According to analyses of collected cases,[31] probably at least 60 per cent of all children with this syndrome are mentally retarded. This may vary from minimal slowing of intellectual function to occasional gross mental deterioration.



Figure 364 Eight year old boy with Sturge-Weber syndrome. Patient is slightly mentally retarded, has numerous seizures, a right hemianopsia and extensive calcification in the left occipital lobe.

Intracortical Calcification: Krabbe[113] in 1934 demonstrated that the intracerebral calcification seen occasionally in this syndrome, most commonly in the occipital lobe on the side of the cutaneous angioma, is in the second and third layers of the cerebral cortex, not in the surface vessels. Microscopically many nerve cells have dropped out and been replaced by glial proliferation. The pia-arachnoid overlying the gyri with calcification shows abundant vascularization but not necessarily angioma formation. On x-ray examination there is a characteristic double-contoured mass of calcification usually in roughly triangular shape within the occipital lobe (Figure 366).

Obesity and sexual retardation are less commonly seen.

The treatment of this condition is not ordinarily surgical. The vascular lesion of the cerebral cortex is a diffuse malformation which does not lend itself to surgical removal and its causal relation to the underlying cerebral atrophy, gliosis and calcification is problematical. Occasionally gross hemorrhage into the subdural space occurs; this has been treated successfully by surgical evacuation (Figure 365).

The main problem in treatment is control of convulsive seizures. This is ordinarily accomplished satisfactorily with dilantin and phenobarbital

in standard doses, or tridione when the attacks are primarily of petit mal type. In this clinic we have not carried out extensive cortical resection for this condition. However, it is possible that in patients with focal seizures of increasing severity, particularly in the presence of hemiplegia and hemianopsia, radical cortical excision, even hemispherectomy, may be the treatment of choice. The possibility of arresting the advance of progressive mental deterioration, as reported after hemispherectomy in other types of infantile hemiplegia must be considered.



Figure 365. Five months old infant with Sturge-Weber syndrome showing cutaneous angioma of the face and neck principally involving the right side. This patient had a subdural hemorrhage on the right which was successfully evacuated. Subsequent development has been fairly satisfactory. Convulsive seizures have been controlled very well with anticonvulsant medication.



Figure 366 Lateral and basal roentgenogram of the skull of a seven year old boy with Sturge-Weber syndrome. Patient had seizures and a right homonymous hemianopsia. Note the characteristic calcification in superficial cortical layers of the left occipital lobe.

PART VIII

EPILEPSY

Chapter I

Epilepsy

SURGICAL TREATMENT of epilepsy is indicated for persistent focal seizures related to a well-localized area of cerebral injury. It has proved of limited application in infants and children because in this age group the lesions which result from cerebral injury are usually diffuse and more widespread than they are in adults. They often involve one whole lobe of the brain or even an entire hemisphere. Recently, it is true, surgical treatment has been extended to radical excision of a whole hemisphere in patients with severe one-sided seizures associated with infantile hemiplegia and progressive mental deterioration due to unilateral cerebral atrophy (Figures 367 and 368).[26, 115]



Figure 367. Antero-posterior, postero-anterior encephalograms of an 11 year old boy with an infantile left hemiplegia. Patient had severe left-sided convulsive seizures Note the extreme atrophy of the entire right hemisphere with the marked shift of mid-line structures from left to right.

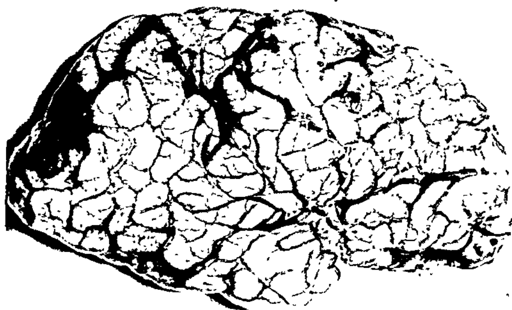


Figure 368. Right cerebral hemisphere removed from 12 year old boy with complete left hemiplegia, 10 to 15 seizures daily on maximal doses of anticonvulsant medication, and progressive mental deterioration. Same patient as encephalograms shown in Figure 367. Post-operative recovery satisfactory and no post-operative seizures to date on no medication.

In infancy and childhood convulsive seizures are extremely common. Almost every illness experienced in this age group, especially those characterized by high fever, is a possible cause for convulsions. The majority of these convulsions are generalized in nature and do not constitute a neurosurgical problem. Their management consists in control of the immediate episode with anticonvulsant sedation and supportive measures together with treatment of the causative disease. Further consideration of this type of convulsion is not within the scope of this chapter.

Focal types of convulsive seizures also may occur in childhood as a result of various inflammatory and toxic processes. They are more apt to occur, however, associated with congenital defects, birth injuries or other local trauma, abscess, porencephaly, or neoplasm. The treatment of convulsions associated with subdural hematoma, brain abscess and brain tumor is essentially the treatment of these primary lesions; they have been dealt with elsewhere. There remains to be considered, then, local cerebral injury due to such congenital defects as local areas of microgyria, brain cysts presumably resulting from fetal or early infantile vascular occlusion, local meningo-cerebral scars or calcification due to birth and neo-natal trauma (Figure 369), and possibly also meningo-cerebral scarring secondary to meningitis.[158]

Certain fundamental concepts regarding focal convulsions should be emphasized before further discussion of surgical treatment is presented.

This is perhaps accomplished best in the words of Penfield and Erickson:[156] "An epileptogenic lesion is a lesion within the central nervous system, which by virtue of its presence, induces in the ganglion cells under its influence a state of hyperactivity, which results in the periodic neuronal discharge of an epileptic seizure. The neurons in or near the pathological lesion which are subject to the irritating influence, form a part of the true, epileptogenic lesion." In other words, a focal convulsive seizure does not arise from a fibrous scar, a cyst, a neoplasm, or an overgrowth of neuroglia, but from a nearby area of cortical gray matter which includes chronically irritated nerve cells. Surgical treatment of focal epilepsy is based on the assumption that clean surgical excision of a cerebral lesion leaves a more benign, non-epileptogenic cortical defect than that which results from a traumatic, vascular, or inflammatory lesion.

The meningo-cerebral lesion which remains as the end result of diffuse pyogenic meningitis is not favorable to surgical treatment. It is too widespread. Although convulsions in this situation may be lateralizing, they are rarely more focal in nature than this.

What are the indications for surgical treatment of convulsive seizures in children? The existence of clinical manifestations of a localized discharge is not in itself sufficient indication for surgery. There should be in addition objective evidence of a well localized pathological process, and the seizures should be of such frequency and severity that they cannot be controlled by



Figure 369 Antero-posterior and lateral encephalograms of 11 year old boy with right-sided seizures showing calcified left frontal mass which does not distort the ventricular system. Pathological diagnosis: gliosis and calcification in cerebral cortex; no evidence of tumor. Seizure-free 10 years following excision.

thorough, supervised trial of reasonable amounts of anticonvulsant medication.

Objective evidence of local pathology is obtained by (1) electroencephalography; (2) pneumoencephalography; (3) arteriography; (4) inspection of the surface of the brain; and (5) electrocorticography, both spontaneous and associated with direct cortical stimulation.

The *electroencephalogram* is almost invariably abnormal in the presence of epilepsy due to local brain injury, whatever its source.[104] Focal disturbances in the electroencephalogram may be two to three per second waves, alone or associated with spikes, bursts of sharp waves, or simply random spikes. These abnormalities are usually constant when due to a local brain injury, but may spread to all leads if a tracing is recorded during a clinical seizure. On the other hand, electroencephalograms which demonstrate generalized dysrhythmia consisting of sharp spikes or spike-and-wave patterns in paroxysms are not apt to be found with local lesions that might be suitable for surgical treatment.



Figure 370. Antero-posterior encephalogram of six year old boy with left hemiplegia and left-sided convulsive seizures. Note extreme atrophy of the right cerebral hemisphere with enlargement of the ventricular and surface subarachnoid pathways. There is a marked shift of the brain toward the atrophic side.

Pneumoencephalography should certainly be carried out on all children with persistent focal seizures in whom surgical treatment is considered a possibility. Such air studies demonstrate ventricular enlargement and irregularity as well as the condition of the cortical subarachnoid pathways. It has long been known that if there is local cortical atrophy or loss of substance, the wall of the ventricle which is nearest migrates toward this area. Thus air studies may show localized protrusion of part of a ventricle toward the surface or diffuse dilatation of an entire lateral ventricle with a shift of the ventricular system toward the dilated side (Figure 370). In addition, cortical atrophy may be indicated by abnormal local collections of air either on the surface or within the hemisphere and connecting with the surface





Figure 371A (*Left*). Antero-posterior roentgenogram showing large porencephalic subcortical cyst of the right hemisphere displacing the ventricular system to the opposite side in infant with numerous left-sided seizures. Treated successfully by establishing communication of this cyst with the basilar cisternae.

B (*Right*). Appearance of the huge subcortical porencephalic cyst exposed at craniotomy.

subarachnoid pathways (porencephaly) (Figure 371). A pneumoencephalogram which demonstrates normal-sized ventricles without distortion or displacement and shows a symmetrical normal pattern of air in the surface pathways virtually rules out the possibility of a cortical lesion suitable for surgical excision in the treatment of epilepsy.

Carotid *arteriography* may demonstrate abnormalities of the cerebral blood vessels giving rise to epileptogenic foci, particularly congenital arteriovenous malformations. The clinical features and treatment of these lesions have been presented elsewhere (p 395).

Inspection of the surface of the cerebral hemisphere at craniotomy may provide in itself the objective evidence which indicates surgical treatment. Lesions which can be identified readily include subdural membranes, vascular malformations, local areas of microgyria, cortical or subcortical cysts, atrophic gyri, adherent meningo-cerebral scars, and, of course, neoplasms.

The most accurate and important objective measurements which help define indications for and extent of cortical excision of epileptogenic cortical foci are provided by: (1) *electrocorticography,* that is, the study of electrical potentials obtained from electrodes placed directly on the cerebral cortex; and by (2) *reproduction of electrocorticographic or clinical seizures* by *electrical stimulation* of the cortex with a current too weak to elicit response from normal brain. Since these studies are most safely and accur-

ately carried out with the patient awake, they are not generally applicable to very young children. A two-stage procedure has been used to advantage in this clinic. In the first operation an osteoplastic bone flap is elevated under general anesthesia in the usual manner; the dura is opened and the cortex inspected. If electrocorticography or stimulation seems to be indicated, the dura and bone flap are replaced. Three to five days later the bone flap is re-elevated under local or very light pentothal anesthesia. When all stimulation and electrocorticography have been completed, general anesthesia is induced for closure.

Cortical Excision

With the cortex exposed under local anesthesia spontaneous electrocorticography is first carried out. By using a special electrode holder clamped to the skull, it is relatively simple to place electrodes at a number of different points on the surface and simultaneously record potentials



Figure 372. Cerebral cortex exposed and multiple leads placed on the brain for electrocorticography prior to cortical resection in the treatment of focal seizures.

between these points and an indifferent electrode (ear), or between various pairs of electrodes (Figure 372). If abnormal foci are not detected spontaneously, low voltage electrical stimulation with a bipolar electrode can be carried out. In children, where reliable cooperation from the patient is apt to be lacking, electrocorticographic demonstration of subclinical seizures in response to cortical stimulation is especially important.

Epileptogenic foci should be sought in the gyri adjacent to visible meningo-cerebral scars, areas of atrophy, local microgyria, or porencephalic cysts. When the epileptogenic focus has been identified, the involved gyri are removed by subpial dissection. An incision is first made in an avascular area and the cortex then removed by gentle suction and blunt dissection down to normal white matter and from sulcus to sulcus. The immediate result of removing such a focus can be checked by replacement of electrodes and restimulation before closure is carried out.

PART IX

LEAD ENCEPHALOPATHY

Chapter I

Lead Encephalopathy

Lead poisoning is of interest to the neurosurgeon because of the high incidence of severe encephalopathy with this disease in the pediatric age group. The disorder is no longer as common as it was more than a decade ago, but it still occurs regularly and new cases have been seen in this clinic every year. By far the commonest source of lead poisoning in infants and young children, at least in this community, is by way of the gastro-intestinal tract. Chewing and sucking paint from toys, furniture, window sills, door jambs and porch railings is apparently a pastime which gives satisfaction to certain infants. Although toys and children's furniture are now generally covered with lead-free paint, other objects about the house may not be. Also, lead poisoning has occurred in children who have chewed the lead-free paint from their cribs only to have these repainted with leaded paint which well-intentioned but misguided parents happened to have in their cellar left over from spring house painting. In another part of the country we have seen lead poisoning result from inhalation of the fumes when

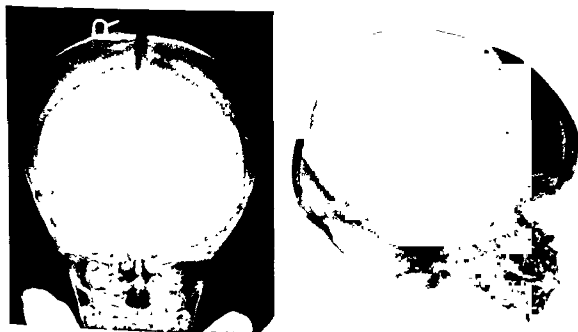


Figure 373. Postero-anterior and lateral roentgenograms of the skull of a two year old infant with extreme intracranial pressure due to lead poisoning Note marked separation of all the cranial sutures with enlargement of the cranial vault.

battery casings containing lead were burned for fuel. Certainly, investigation of undiagnosed coma, convulsions, increased intracranial pressure or other evidence of central nervous system disease in childhood should include careful inquiry into the possibility of ingestion or inhalation of lead.

Clinical Features

Although ingestion of lead may occur in small amounts over fairly long periods of time, the onset of encephalopathy may be sudden and severe. In children, symptoms referable to the nervous system are often the first evidence of lead toxicity. Marked weakness, lethargy and anorexia are soon followed by generalized convulsions and symptoms and signs of increased intracranial pressure. Convulsions are often unusually violent requiring

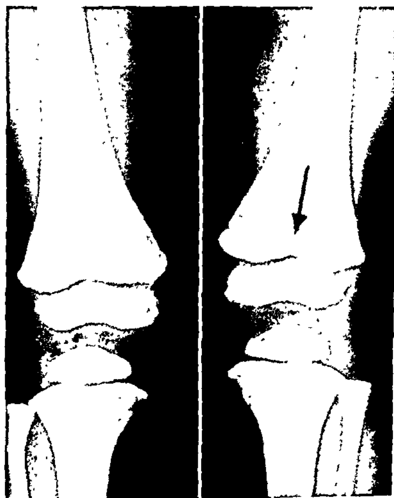


Figure 374 Characteristic x-ray appearance of the long bones in severe lead poisoning as seen in an antero-posterior roentgenogram of the knees. *There is a wide band of increased density* just beneath the epiphyseal cartilage This consists of parallel trabeculae composed chiefly of calcified intracellular substance containing large quantities of lead. Patient was a 2 4/12 year old girl.

general anesthesia for control. The child is apt to be generally hypertonic and either highly irritable or stuporous. Vomiting is frequent. In infants there is bulging of the anterior fontanelle, dilatation of the scalp veins and splitting of the cranial sutures. There is usually marked papilledema with many fresh hemorrhages in the retina.

Lumbar puncture reveals clear cerebrospinal fluid under markedly increased pressure. The cell count is abnormal, varying from eight or 10 up to as many as *100 or more lymphocytes* per cubic millimeter. The protein content is always elevated and the degree of this increase seems to be in proportion to the severity of the encephalopathy.

X-ray findings include separation of the cranial sutures (Figure 373) and a lead line in the long bones. The latter consists of a zone of increased density adjacent to the epiphyseal cartilage which is best seen perhaps in an anteroposterior film of the knees (Figure 374). Here the lead line is visualized at the lower end of the femur and the upper end of the tibia and fibula.

Laboratory studies may be of interest in demonstrating an anemia with increase in immature types of red blood cells, many of which show basophilic stippling. Twenty-four hour urine samples may show significant amounts of excreted lead.



Figure 375 (*Upper*). Severe lead encepha- ... A large fronto- ... been removed. ... the tremendous *increase in intracranial pressure causing the* dura to be bloodless and almost transparent.

Figure 376 (*Lower*) Same patient shown in Figure 375 with lead encephalopathy after opening of the dura. There is marked venous *congestion and generalized edema of the brain.*

Morbid Anatomy

In severe lead encephalopathy the brain is diffusely swollen to an almost unbelievable degree. There is marked increase in the weight of the brain, flattening and pallor of the convolutions, and downward

displacement at the incisura of the tentorium and at the foramen magnum. This is presumably due to tremendous increase in interstitial fluid resulting from generalized capillary wall damage. The ventricles and surface subarachnoid pathways are compressed and therefore smaller than normal.

Treatment

The morbidity and mortality of lead encephalopathy in childhood are very high unless vigorous treatment is carried out with dispatch early in the course of the disease. The immediate problems are control of convulsive seizures and reduction of increased intracranial pressure before irreversible changes occur. In mild degrees of encephalopathy it may be possible to control seizures with standard doses of anticonvulsants and to control intracranial pressure with frequent lumbar punctures or constant spinal or ventricular drainage supplemented by repeated regular use of hypertonic solutions such as 50 per cent glucose, magnesium sulphate, concentrated plasma, or albumin. At the same time efforts are made to promote deposition of lead in the bones.

In more severe degrees of acute lead poisoning, these measures will be inadequate to reduce the extreme degree of increased intracranial pressure and surgical decompression should be carried out. This is indicated when-

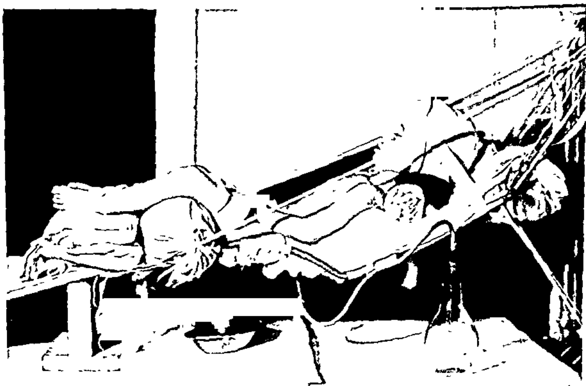


Figure 377 Patient on constant spinal drainage after bilateral cranial decompression for severe lead encephalopathy with extreme increased intracranial pressure. Lateral ventricles may be so small that constant ventricular drainage is impossible to maintain.

ever the vital signs remain embarrassed, convulsions cannot be controlled, the protein content of the spinal fluid continues rising, or increasing depth of coma is present. An ordinary subtemporal decompression is of little avail. It is the practice in this clinic, if surgical treatment is decided upon, to perform at once as extensive a decompression as possible. A large fronto-parieto-temporal scalp flap is raised followed by elevation and removal of an equally large bone flap. The dura is usually pale and extremely tense (Figure 375); it should be opened widely as quickly as possible to prevent rupture of the brain through a small incision (Figure 376). The dura is left open to the limits of the bony exposure and the brain is covered with a piece of polyethylene film. The scalp is then resutured tightly. The same procedure is usually carried out on both sides at the same sitting. The bone flaps are boiled and preserved for reinsertion subsequently if the outcome is favorable. Spinal fluid drainage and administration of hypertonic solutions should be used to supplement surgical decompression (Figure 377).



Figure 378. Eleven year old girl approximately nine years after bilateral removal of fronto-temporo-parietal bone flaps for severe lead encephalopathy with extreme increased intracranial pressure which resulted in deep coma for almost three weeks. Excellent recovery of function and progressive mental development.

There is always some question as to whether this heroic type of treatment is wise because, if it is necessary, the probability is great that severe brain damage has already occurred. However, there is no way to be certain of this in the acute phase of severe lead encephalopathy and surgical decompression may unquestionably sometimes be a life-saving maneuver. The prognosis for life and normal mentality after this degree of encephalopathy is poor but by no means hopeless (Figure 378).

PART X

PEDIATRIC NEUROSURGICAL ANESTHESIA

Chapter I

Pediatric Neurosurgical Anesthesia

THE SMOOTHNESS with which the average intracranial operation proceeds in infancy and childhood today should in large part be credited to advances in anesthesiology. The anesthetic agent of choice in our clinic is ether administered by endotracheal insufflation through a Y tube (Figure 379A). Intubation is now employed for every inhalation anesthetic administration in this clinic except perhaps in very short procedures when light anesthesia only is indicated. Certainly in all operations about the head and neck where the nose and mouth are difficult of access to the anesthetist, it is highly desirable. It is considered essential for every inhalation anesthesia administered to a child who is to be in the prone position during operation, as for posterior fossa exploration or cervical or upper thoracic laminectomy. Age is no contra-indication to endotracheal intubation; it may be employed in the first year of life just as satisfactorily as in older children if expertly performed. For children of all ages, even more than in adults, it is imperative that endotracheal tubes be kept scrupulously clean.

In children with posterior fossa neoplasms or with internal hydrocephalus due to obstruction to spinal fluid circulation from any cause, intracranial manipulations are apt to be accompanied by respiratory irregularity or depression, or even sudden complete apnea. The advantages of endotracheal intubation under these circumstances in permitting artificial respiration until spontaneous breathing is resumed are obvious. If there has been unusual difficulty in performing intubation so that edema of the vocal cords or larynx is anticipated, it may be wise to place a child in a humid atmosphere (steam tent or room) after extubation until an adequate airway is assured.

Induction of anesthesia in the frightened uncooperative child may be an important psychological as well as physiological problem.[153] A minimum of unfamiliar and unpleasant odors and tastes which lead to anxiety, resistance and voluntary breath-holding is desirable. Hypoxia, of course, should be avoided during induction, particularly in patients with increased intracranial pressure. In this clinic, rectal avertin (tribromethanol) in doses of 80 mg./kilo. of body weight has proved to be the most satisfactory basal analgesic for neurosurgical patients over one year of age. Its use is contra-indicated only in the presence of liver disease, profound anemia, or

an incompetent rectal sphincter. Avertin is made up in a 2.5 per cent solution and administered through a soft rubber catheter. A cleansing enema must be part of the pre-operative preparation. It may be wise to use a Foley type catheter or to hold the buttocks together firmly after introduction of the avertin to prevent its expulsion before absorption has occurred. If avertin is employed, it is our custom to give no additional pre-operative medication except scopolamine or atropine. In particular, morphine should be avoided because it may cause undue depression when combined with avertin.

In infants under one year of age, the threshold for pain is high and psychological factors are minimal. It is therefore usually unnecessary to give any pre-anesthetic sedation in this age group. In older children who are apprehensive and in good general health, especially those who do not have increased intracranial pressure, pre-medication with morphine, demerol or a barbiturate followed by nitrous oxide and oxygen, pentothal, or divinyl ether (vinethene) induction may be preferable to avertin. Amounts of these sedatives recommended for average, well-developed patients are shown in Table XII. In chronically ill or malnourished patients, these doses

TABLE XII

PRE-OPERATIVE MEDICATION FOR INFANTS AND CHILDREN

Age	*Average Weight*	*Seconal or Nembutal*	*Morphine*	*Scopolamine or Atropine*	*Demerol*
New-born	7 lbs.	—	—	0 1 mg	—
6 months	16 lbs	30 mg	—	0.2 mg.	—
1 year	21 lbs	50 mg.	1.0 mg.	0.2 mg.	10 mg.
2 years	27 lbs.	60 mg.	1.4 mg.	0.3 mg.	20 mg.
4 years	35 lbs.	90 mg.	2.4 mg	0 3 mg.	25 mg
6 years	45 lbs	100 mg	4 0 mg.	0 3 mg	40 mg.
8 years	55 lbs	120 mg	5 4 mg.	0 4 mg	45 mg.
10 years	65 lbs	150 mg.	6 0 mg.	0.4 mg.	50 mg.
12 years	80 lbs	200 mg	8 0 mg.	0 6 mg	50 mg

should be reduced. All infants and children who are scheduled to have an inhalation anesthesia should receive scopolamine or atropine to minimize oral and nasopharyngeal secretions and to block vagal reflexes. Average doses are shown in Table XII. Barbiturates should be administered subcutaneously or rectally 60 to 90 minutes before the scheduled time for operation; morphine, demerol and scopolamine should be administered subcutaneously 45 to 60 minutes before operation, or intravenously when immediate effect is desired.

In infants and small children, the period of induction is the most critical

and most in need of expert supervision. The transitions between voluntary breath-holding, involuntary laryngeal spasm and apnea due to profound anesthesia may be very abrupt in this age group. If there is marked increase in intracranial pressure, as in internal hydrocephalus due to posterior fossa obstruction, the pressure should be released by ventricular puncture prior to beginning the induction. Children who are to be operated upon in the prone position should not be turned to this position until anesthesia has been satisfactorily induced and is proceeding smoothly during a short period of observation.

Pentothal, curare, and the various intraspinal agents have had little place in pediatric anesthesia in this clinic. As an inducing agent only in healthy older children with no pre-operative respiratory irregularity or depression, pentothal may be used carefully. It can be administered as a 0.6 per cent solution per rectum (1.-1.5 ccs. per pound of body weight) or as a 2.5 per cent solution intravenously, usually in amounts of 2. to 5. ccs., depending on the size of the child.

Local anesthesia in our clinic has been reserved almost entirely for short superficial operative procedures on premature or new-born babies. It has proved satisfactory for elevation of depressed skull fractures and repair of small encephaloceles or spinal meningoceles. In this group of patients it is supplemented to advantage by use of a sugar nipple reinforced with a few drops of paregoric or whisky. Occasionally in older children who are moribund or debilitated, local anesthesia has been used alone or in combination with avertin administered rectally. Local anesthesia alone or in combination with avertin or barbiturates is also indicated in certain diagnostic procedures such as myelography or ventriculography in older children.

It is well to emphasize, however, that in our experience the safest and most satisfactory anesthesia for small infants, for children in poor nutritional states, and for children critically ill from increased intracranial pressure, malignancy or infection is ethyl ether administered with adequate amounts of oxygen.

In addition to expertly administered anesthesia, young children need adequate parenteral hydration and control of body temperature in order to tolerate long operative procedures. An inlying cannula or plastic catheter for constant infusion of fluids,[3] blood or supportive medications is a prerequisite to every major neurosurgical procedure in childhood (Figure 379D). Facilities should be available to pump blood into the patient rapidly during periods of active bleeding at the operative site (Figure 379E). It is well to remember that 25 ccs. blood loss in a new-born infant is equal to a 500 to 600 ccs. hemorrhage in an adult patient. Methods of controlling temperature during operation have been discussed elsewhere

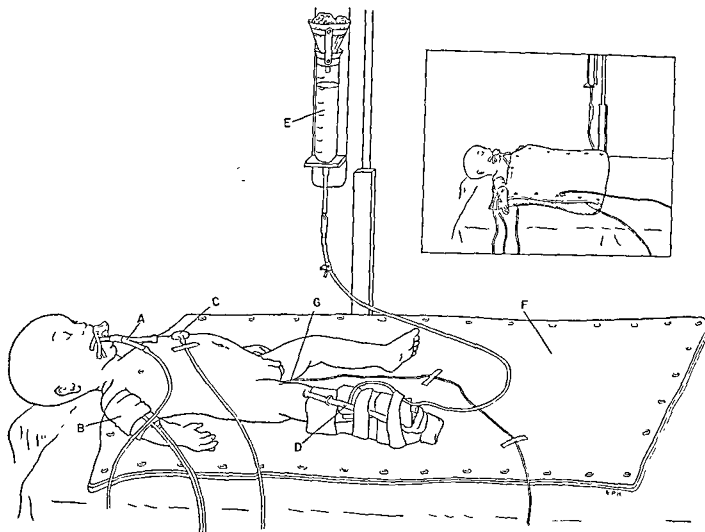


Figure 379. Infant on operating table in position for frontal craniotomy.

A. Endotracheal tube with Y connection for controlled administration of inhalation anesthesia.

B. Blood pressure cuff on right arm with accompanying stethoscope strapped to the antecubital fossa.

C. Stethoscope strapped over the cardiac apex.

D. Plastic catheter placed in saphenous vein in the ankle and attached by means of side-arm syringe to constant intravenous drip.

E. Reservoir of intravenous fluid or blood to be administered throughout operation. Three-way stop-cock in the intravenous tubing permits rapid pumping of infusion or intravenous injection of stimulants as necessary.

F Insulated electric blanket which is thermostatically controlled.

G. Thermocouple in baby's rectum attached by insulated wire to potentiometer on the operating room wall for constant recording of baby's temperature throughout operation Insert shows the electric blanket folded over the baby's trunk and lower extremities

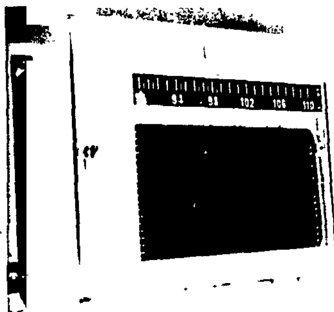


Figure 380. Potentiometer on the wall of the operating room recording the patient's rectal temperature constantly throughout operation. Lead-in wire from thermocouple in baby's rectum is seen in the lower right hand corner.

(Figure 379F, G). Since changes in temperature, pulse, respirations and blood pressure occur so rapidly in infants and children, they should always be anticipated and measures to counteract them begun as early as possible (Figure 380).

Following long operative procedures of a critical nature, particularly posterior fossa tumors, it is often wise to leave the child on the operating table for some time. It may be a disturbing experience to move such patients too quickly, and if they are allowed to stabilize temperature, pulse and blood pressure and if possible to recover consciousness or at least cough and swallowing reflexes before being transported, post-operative recovery is usually greatly facilitated.

Local topical anesthesia for the introduction of needles through the skin may be of considerable value in young children, particularly when repeated subcutaneous or intramuscular injections are necessary over long periods of time. This has been accomplished successfully by the local application of cold – a metallic surface cooled to 4° to 6° C. and applied to the skin surface for 45 to 60 seconds.[100] A compact unit using freon gas in cartridges (Figure 381) is being developed now for routine use. If a small area of skin is cooled in this manner down to 11° to 13° C., hypodermic needles can be introduced entirely without pain and with virtually no sensation at all if the pressure of introduction of the needle is equalized by counter pressure on the adjacent skin.

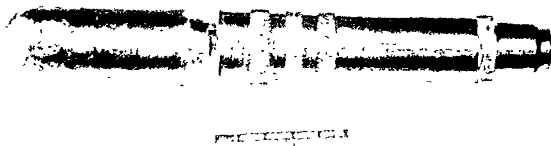


Figure 381. Cooling unit for production of transient cutaneous anesthesia for introduction of hypodermic needles in infants and children. The flat surface at the right end of the instrument is cooled to 4° to 6° C. by the release of freon gas contained in the replaceable cartridge at the left end of instrument.

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This Book

NEUROSURGERY
of Infancy and Childhood

By

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and
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